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RESEARCH, TREATMENT, AND PREVENTION OF
SICKLE CELL ANEMIA

GOVERNMENT DOCUMENTS

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HEARING

BEFORE THE

KANSAS STATE UNIVERSITY

SUBCOMMITTEE ON

PUBLIC HEALTH AND ENVIRONMENT

OF THE

COMMITTEE ON

INTERSTATE AND FOREIGN COMMERCE

HOUSE OF REPRESENTATIVES

NINETY-SECOND CONGRESS

FIRST SESSION

ON

H.R. 11742, H.R. 7654, and H.R. 11171
(and all identical bills)

BILLS TO PROVIDE FOR THE PREVENTION OF SICKLE CELL
ANEMIA AND TO AMEND THE PUBLIC HEALTH SERVICE ACT
TO PROVIDE FOR THE ESTABLISHMENT OF A NATIONAL
SICKLE CELL ANEMIA INSTITUTE

NOVEMBER 12, 1971

Serial No. 92-57

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RESEARCH, TREATMENT, AND PREVENTION OF SICKLE CELL ANEMIA

FRIDAY, NOVEMBER 12, 1971

HOUSE OF REPRESENTATIVES,
SUBCOMMITTEE ON PUBLIC HEALTH AND ENVIRONMENT,
COMMITTEE ON INTERSTATE AND FOREIGN COMMERCE,
Washington, D.C.

The subcommittee met at 10 a.m., pursuant to notice, in room 2318, Rayburn House Office Building, Hon. Paul G. Rogers (chairman) presiding.

Mr. ROGERS. The subcommittee will come to order, please.

Today, we are opening hearings to determine the necessity of accelerated programs of research, treatment, and prevention of sickle cell anemia.

It is generally agreed that 40,000 to 50,000 persons in the United States have this disease and an additional 8 to 13 percent of the black population carry the trait. The disease is said to occur in approximately 1 in 500 black births in the United States, and median survival age for victims of sickle cell anemia is 20 years.

There is no known cure for this disease since it is caused genetically, and treatment methods presently used do not appear to provide complete relief and may be accompanied by serious side effects.

A number of tests are available for the detection and diagnosis of sickle cell disease. Coupled with voluntary genetic counseling services, these tests can be used as a means toward preventing unwanted occurrence of the disease.

Federal research support in sickle cell anemia is being provided at the National Institutes of Health and is coordinated by the National Heart and Lung Institute. During fiscal year 1971 this effort was funded at approximately \$1 million.

This year an additional \$5 million was appropriated by Congress at the request of the President for expanded programs in sickle cell anemia, increasing the amount of money to be allocated for sickle cell research for this fiscal year to \$6 million. It is the committee's understanding that a Sickle Cell Advisory Committee has been established within HEW to recommend means of carrying out the expanded sickle cell program, but that none of the additional \$5 million has been spent to date.

The purpose of these hearings is to determine the adequacy of the present research and treatment effort and to develop fully the plans of the National Institutes of Health for the expanded program. We are also considering various bills which would increase the Federal effort toward combating this badly neglected disease. H.R. 7654, introduced by Mrs. Grasso, and several identical bills would authorize the establishment of a separate National Sickle Cell Institute. H.R. 11171, in-

roduced by Mr. Fauntroy and others, as well as a number of identical bills, would make available \$25 million a year for the establishment of screening and counseling programs and a further \$5 million a year for research into treatment and diagnosis of the disease.

I and several of my colleagues on the committee (Mr. Kyros, Mr. Preyer, Mr. Symington, Mr. Roy, Mr. Nelsen, Mr. Carter, and Mr. Hastings) have introduced legislation similar to this bill, but without specific authorization figures.

(The text of the bills referred to and agency reports thereon follow:)

92D CONGRESS
1ST SESSION

H. R. 11742

IN THE HOUSE OF REPRESENTATIVES

NOVEMBER 11, 1971

Mr. ROGERS (for himself, Mr. KYROS, Mr. PREYER of North Carolina, Mr. SYMINGTON, Mr. ROY, Mr. NELSEN, Mr. CARTER, and Mr. HASTINGS) introduced the following bill; which was referred to the Committee on Interstate and Foreign Commerce

A BILL

To provide for the prevention of sickle cell anemia.

1 *Be it enacted by the Senate and House of Representa-*
2 *tives of the United States of America in Congress assembled,*

3 SHORT TITLE

4 SECTION 1. This Act shall be cited as the "National
5 Sickle Cell Anemia Prevention Act".

6 FINDINGS AND DECLARATION OF PURPOSE

7 SEC. 2. (a) The Congress hereby finds and declares—

8 (1) that sickle cell anemia is a disease resulting
9 from the inheritance of a genetic factor relating to the
10 sickle cell trait which afflicts a large number of Ameri-
11 can citizens, primarily among the black population of the
12 United States;

1 (2) that the disease is a deadly and tragic burden
2 which strikes approximately one of every five hundred
3 black children, and that less than half of those children
4 who contract the disease survive beyond the age of
5 twenty; and

6 (3) that efforts to prevent sickle cell anemia must
7 be directed toward increased research in the cause and
8 treatment of the disease, and the education, screening,
9 and counseling of carriers of the sickle cell trait;

10 (4) that simple and inexpensive screening tests
11 have been devised which will identify those who have
12 the disease or carry the trait;

13 (5) that programs to prevent sickle cell anemia
14 must be based entirely upon the voluntary cooperation
15 of the individuals involved;

16 (6) that the attainment of better methods of pre-
17 vention, diagnosis, and treatment of sickle cell anemia
18 deserves the highest priority.

19 (b) In order to preserve and protect the health and wel-
20 fare of all citizens, it is the purpose of this Act to establish
21 a national program for the prevention and treatment of
22 sickle cell anemia.

23 AMENDMENTS TO PUBLIC HEALTH SERVICE ACT

24 SEC. 3. (a) Section 1 of the Public Health Service Act
25 is amended by striking out "titles I to X" and inserting in
26 lieu thereof "titles I to XI".

1 (b) The Act of July 1, 1944 (58 Stat. 682), as
2 amended, is amended by renumbering title XI (as in effect
3 prior to the enactment of this Act) as title XII, and by
4 renumbering sections 1101 through 1114 (as in effect prior
5 to the enactment of this Act), and references thereto, as
6 sections 1201 through 1214, respectively.

7 (c) The Public Health Service Act is further amended
8 by adding after title X the following new title:

9 "TITLE XI—SICKLE CELL ANEMIA PREVENTION
10 PROGRAM

11 "GRANTS FOR SICKLE CELL SCREENING AND
12 COUNSELING PROGRAMS

13 "SEC. 1101. (a) The Secretary is authorized to make
14 grants to and enter into contracts with public and non-
15 profit private entities to assist in the establishment and op-
16 eration of voluntary sickle cell anemia screening and coun-
17 seling programs and to assist in developing and making
18 available information and educational materials relating to
19 sickle cell anemia to all persons requesting such information
20 or materials, and to inform the public generally about the
21 nature of sickle cell anemia and the sickle cell trait.

22 "(b) In making grants and contracts under this section
23 the Secretary shall take into account the number of persons
24 to be served, the extent to which such screening and coun-
25 seling is needed on a local basis, the relative need of the

1 applicant, and its capacity to make rapid and effective use of
2 such assistance.

3 “(c) For the purpose of making payments pursuant to
4 grants and contracts under this section, there are authorized
5 to be appropriated such sums as may be necessary for the
6 fiscal year ending June 30, 1973, and the two succeeding
7 fiscal years.

8 “DEMONSTRATION GRANTS

9 “SEC. 1102. (a) In order to promote research in the
10 diagnosis, treatment, and prevention of sickle cell anemia,
11 development of programs to educate the public regarding the
12 nature and inheritance of the sickle cell trait and sickle cell
13 anemia, and the development of centers for research, test-
14 ing, counseling, prevention, or treatment of sickle cell
15 anemia, the Secretary is authorized to make grants to public
16 or nonprofit private entities and to enter into contracts with
17 public or private entities and individuals for projects for
18 research and research training in such fields.

19 “(b) For the purpose of making payments pursuant
20 to grants and contracts under this section there are authorized
21 to be appropriated such sums as may be necessary for the
22 fiscal year ending June 30, 1973, and the two succeeding
23 fiscal years.

24 “VOLUNTARY PARTICIPATION

25 “SEC. 1103. The participation by any individual in

1 any program or portion thereof under this title (whether
2 by grant or contract) shall be wholly voluntary and shall
3 not be a prerequisite to eligibility for or receipt of any other
4 service or assistance from or to participation in any other
5 program.

6 "APPLICATIONS

7 "SEC. 1104. A grant under this title may be made
8 upon application to the Secretary at such time, in such
9 manner, containing and accompanied by such information as
10 the Secretary deems necessary. Each application shall—

11 "(1) provide that the programs and activities for
12 which assistance under this title is sought will be ad-
13 ministered by or under the supervision of the applicant;

14 "(2) describe with particularity the programs and
15 activities for which assistance is sought;

16 "(3) provide for strict confidentiality of all test
17 results, medical records and other information regarding
18 screening, counseling, or treatment of any person
19 treated, except for (A) such information as the patient
20 (or his guardian) consents to be released; or (B) sta-
21 tistical data compiled without reference to the identity of
22 any such patient;

23 "(4) provide for appropriate consultation with com-
24 munity representatives in the development and operation
25 of any program funded under this title;

1 “(5) set forth such fiscal control and fund ac-
2 counting procedures as may be necessary to assure
3 proper disbursement of and accounting for Federal funds
4 paid to the applicant under this title; and

5 “(6) provide for making such reports in such
6 form and containing such information as the Secretary
7 may reasonably require.

8 “PUBLIC HEALTH SERVICE FACILITIES

9 “SEC. 1105. The Secretary shall establish a program
10 within the Public Health Service to provide for voluntary
11 sickle cell anemia screening, counseling, and treatment. Such
12 program shall be made available through facilities of the
13 Public Health Service to any eligible person requesting
14 screening, counseling, or treatment, and shall include notifi-
15 cation of all eligible persons of the availability and voluntary
16 nature of such programs.

17 “REPORTS

18 “SEC. 1106. (a) The Secretary shall prepare and sub-
19 mit to the President for transmittal to the Congress on or
20 before April 1 of each year a comprehensive report on the
21 administration of this title.

22 “(b) The report required by this section shall contain
23 such recommendations for additional legislation as the Sec-
24 retary deems necessary.”

PROTECTION OF ARMED FORCES PERSONNEL

SEC. 4. (a) The Secretary of Defense is authorized and directed to promulgate rules and regulations to provide for screening and counseling of members of the Armed Forces (including their dependents), civilian employees of the Department of Defense, and persons examined at Armed Forces examining and entrance stations, for the sickle cell trait and sickle cell anemia.

(b) Such rules and regulations shall provide for—

(1) voluntary screening for the sickle cell trait for persons described in subsection (a) who request such a test, at no cost to such person;

(2) communication to such person described in subsection (a) of the results of such test;

(3) voluntary referral of individuals determined to possess a positive trait to an appropriate military or civilian counseling or treatment agency;

(4) notification to persons described in subsection (a) of the cost-free and voluntary nature of the screening and referral programs implemented pursuant to this section;

(5) education of persons described in subsection (a) regarding the nature and inheritance of the sickle cell trait and sickle cell anemia; and

1 (6) assurance that all information obtained on
2 specimens submitted voluntarily under this Act shall be
3 held confidential except for (A) such information as the
4 patient (or his guardian) consents to be released or (B)
5 statistical data compiled without reference to the iden-
6 tity of any such patient.

7 (c) The Secretary of Defense shall provide for volun-
8 tary counseling or treatment of such persons described in
9 subsection (a) found to have the sickle cell trait or sickle
10 cell anemia at an appropriate military or civilian facility as
11 the case may be.

12 (d) (1) The Secretary of Defense shall prepare and
13 submit to the President for transmittal to Congress on or
14 before April 1 of each year a comprehensive report on the
15 administration of this section.

16 (2) The report required by this subsection shall con-
17 tain such recommendations for additional legislation as the
18 Secretary of Defense deems necessary.

19 (e) The participation by any individual in any pro-
20 gram or portion thereof under this section shall be wholly
21 voluntary and shall not be a prerequisite to eligibility for or
22 receipt of any other service or assistance from, or to partici-
23 pation in, any other program.

PROTECTION OF VETERANS

SEC. 5. (a) Chapter 17 of title 38, United States Code, is amended by adding at the end thereof the following new subchapter:

**"SUBCHAPTER VI—SICKLE CELL ANEMIA
PREVENTION**

"§ 651. Notification and education

"(a) The Administrator shall notify all persons eligible for care under this chapter of the availability of screening, treatment, and counseling programs with regard to the sickle cell trait and sickle cell anemia and the voluntary nature of such programs.

"(b) The Administrator shall establish a program of education regarding the nature and inheritance of sickle cell trait and sickle cell anemia and make such program available to all such eligible persons.

"§ 652. Screening and treatment

"(a) The Administrator shall furnish to any person eligible for care under this chapter who makes a request, screening for sickle cell trait or sickle cell anemia.

"(b) Upon a finding that such eligible person has the sickle cell trait or sickle cell anemia the Administrator shall provide for voluntary counseling or treatment as the case may be.

1 **“§ 653. Reports**

2 “(a) The Administrator shall prepare and submit to the
3 President for transmittal to the Congress on or before April 1
4 of each year a comprehensive report on the administration
5 of this subchapter.

6 “(b) The report required by this section shall contain
7 such recommendations for additional legislation as the Ad-
8 ministrator deems necessary.

9 **“§ 654. Voluntary participation**

10 “The participation by any individual in any program
11 or portion thereof under this section shall be wholly volun-
12 tary and shall not be a prerequisite to eligibility for or
13 receipt of any other service or assistance from, or to partici-
14 pation in, any other program.”

15 (b) The analysis at the beginning of such chapter is
16 amended by adding at the end thereof:

“SUBCHAPTER VI—SICKLE CELL ANEMIA PREVENTION

“651. Notification and education.

“652. Screening and treatment.

“653. Reports.

“654. Voluntary participation.”

[H.R. 7654, 92d Cong., 1st sess., introduced by Mrs. Grasso on April 22, 1971; H.R. 8423, 92d Cong., 1st sess., introduced by Mrs. Hicks of Massachusetts on May 17, 1971;

H.R. 8597, 92d Cong., 1st sess., introduced by Mrs. Grasso (for herself, Mr. Addabbo, Mr. Aspin, Mr. Badillo, Mr. Brasco, Mr. Burke of Massachusetts, Mr. Burton, Mr. Collins of Illinois, Mr. Conyers, Mr. Edwards of California, Mr. Eilberg, Mr. Fauntroy, Mr. Halpern, Mr. Hawkins, Mr. Hechler of West Virginia, Mr. Horton, Mr. McKinney, and Mr. Cotter) on May 20, 1971;

H.R. 8598, 92d Cong., 1st sess., introduced by Mrs. Grasso (for herself, Mr. Metcalfe, Mr. Miller of California, Mr. Mitchell, Mr. Moss, Mr. Murphy of New York, Mr. Pepper, Mr. Rangel, Mr. Riegle, Mr. Reuss, Mr. Roe, Mr. Rosenthal, Mr. Ryan, Mr. Sarbanes, Mr. Steele, Mr. Stokes, and Mr. Charles H. Wilson) on May 20, 1971;

H.R. 10791, 92d Cong., 1st sess., introduced by Mr. O'Neill on September 21, 1971;

H.R. 10932, 92d Cong., 1st sess., introduced by Mr. Rangel on September 28, 1971; and

H.R. 11004, 92d Cong., 1st sess., introduced by Mr. Moorhead on September 30, 1971, are identical as follows:]

A BILL

To amend the Public Health Service Act to provide for the establishment of a National Sickle Cell Anemia Institute.

1 *Be it enacted by the Senate and House of Representa-*
 2 *tives of the United States of America in Congress assembled,*
 3 That title IV of the Public Health Service Act (42 U.S.C.,
 4 ch. 6A, subch. III) is amended by adding at the end
 5 thereof the following new part:

6 "PART G.—NATIONAL SICKLE CELL ANEMIA INSTITUTE

7 "ESTABLISHMENT OF NATIONAL SICKLE CELL ANEMIA

8 INSTITUTE

9 "SEC. 461. For the purpose of conducting and support-
 10 ing programs for the diagnosis, treatment, and prevention of
 11 sickle cell anemia, the Secretary shall establish in the Public

I

1 Health Service an institute to be known as the National
2 Sickle Cell Anemia Institute (hereafter in this part referred
3 to as the 'Institute').

4 "FUNCTIONS

5 "SEC. 462. (a) The Secretary, through the Institute,
6 shall—

7 "(1) conduct and support (through grants or con-
8 tracts) research programs for the diagnosis, treatment,
9 and prevention of sickle cell anemia.

10 "(2) provide training and instruction and establish
11 traineeships and fellowships, in the Institute and else-
12 where, in matters relating to the diagnosis, treatment,
13 and prevention of sickle cell anemia,

14 "(3) provide for nationwide screening programs to
15 determine the incidence of sickle cell anemia and its
16 traits among school age children,

17 "(4) conduct and support (through grants or con-
18 tracts) counseling and education programs, developed
19 in consultation with community representatives, to make
20 individuals and communities aware of the services avail-
21 able with respect to sickle cell anemia and to make
22 individuals aware of their chances of carrying the dis-
23 ease, and

24 "(5) assist the Secretary of Defense in screening all

1 persons entering the armed forces for sickle cell anemia
2 and its traits.

3 “(b) The Secretary, through the Institute, shall also
4 carry out the purposes of section 301 with respect to re-
5 search, investigations, experiments, demonstrations, and
6 studies related to sickle cell anemia, except that the Sec-
7 retary shall determine the areas in which and the extent
8 to which he will carry out such purposes of section 301
9 through the Institute or another institute established by or
10 under other provisions of this Act, or both of them, when
11 both such institutes have functions with respect to the same
12 subject matter.

13 “(c) The Secretary may provide trainees and fellows
14 participating in training and instruction or in traineeships and
15 fellowships under subsection (a) (2) with such stipends and
16 allowances (including travel and subsistence expenses) as
17 he deems necessary, and, in addition, provide for such train-
18 ing, instruction, traineeships, and fellowships through grants
19 to public or other nonprofit institutions.

20 “ESTABLISHMENT OF ADVISORY COUNCIL

21 “SEC. 463. (a) The Secretary shall establish an ad-
22 visory council to advise, consult with, and make recom-
23 mendations to, him on matters relating to the Institute.

24 “(b) The provisions relating to the composition, terms

1 of office of members, and reappointment of members of
2 advisory councils under section 432 (a) shall be applicable
3 to the advisory council established under this section, ex-
4 cept that the Secretary may include on such advisory council
5 such additional ex-officio members as he deems necessary.
6 “(c) Upon appointment of such advisory council, it
7 shall assume all, or such part as the Secretary may specify,
8 of the duties, functions, and powers of the National Ad-
9 visory Health Council relating to the research or training
10 projects with which the advisory council established under
11 this part is concerned and such portion as the Secretary
12 may specify of the duties, functions, and powers of any
13 other advisory council established under this Act relating
14 to such projects.”

[H.R. 11171, 92d Cong., 1st sess., introduced by Mr. Fauntroy (for himself, Mrs. Chisholm, Mr. Clay, Mr. Collins of Illinois, Mr. Conyers, Mr. Dellums, Mr. Diggs, Mr. Fraser, Mr. Hawkins, Mr. Metcalfe, Mr. Mitchell, Mr. Rangel, and Mr. Stokes) on October 12, 1971;

H.R. 11257, 92d Cong., 1st sess., introduced by Mr. Widnall on October 14, 1971; H.R. 11420, 92d Cong., 1st sess., introduced by Mr. Green of Pennsylvania (for himself, Mrs. Chisholm, Mr. Collins of Illinois, Mr. Dellums, Mr. Diggs, Mr. Fauntroy, Mr. Fraser, Mr. Hawkins, Mr. Mikva, and Mr. Mitchell) on October 26, 1971; and

H.R. 11421, 92d Cong., 1st sess., introduced by Mr. Anderson of California on October 27, 1971, are identical as follows:]

A BILL

To provide for the prevention of sickle cell anemia.

1 *Be it enacted by the Senate and House of Representa-*
2 *tives of the United States of America in Congress assembled,*

3 SHORT TITLE

4 SECTION 1. This Act shall be cited as the "National
5 Sickle Cell Anemia Prevention Act".

6 FINDINGS AND DECLARATION OF PURPOSE

7 SEC. 2. (a) The Congress hereby finds and declares—

8 (1) that sickle cell anemia is a disease resulting
9 from the inheritance of a genetic factor relating to the
10 sickle cell trait which afflicts a large number of Amer-
11 ican citizens, primarily among the black population of
12 the United States;

1 (2) that the disease is a deadly and tragic burden
2 which strikes approximately one of every five hundred
3 black children, and that less than half of those children
4 who contract the disease survive beyond the age of
5 twenty; and

6 (3) that efforts to prevent sickle cell anemia must
7 be directed toward increased research in the cause and
8 treatment of the disease, and the education, screening, and
9 counseling of carriers of the sickle cell trait;

10 (4) that simple and inexpensive screening tests
11 have been devised which will identify those who have
12 the disease or carry the trait;

13 (5) that programs to prevent sickle cell anemia
14 must be based entirely upon the voluntary cooperation
15 of the individuals involved; and

16 (6) that the attainment of better methods of pre-
17 vention, diagnosis, and treatment of sickle cell anemia
18 deserve the highest priority.

19 (b) In order to preserve and protect the health and
20 welfare of all citizens, it is the purpose of this Act to estab-
21 lish a national program for the prevention and treatment of
22 sickle cell anemia.

23 AMENDMENTS TO PUBLIC HEALTH SERVICE ACT

24 SEC. 3. (a) Section 1 of the Public Health Service Act
25 is amended by striking out "Titles I to X" and inserting in
26 lieu thereof "Titles I to XI".

1 (b) The Act of July 1, 1944 (58 Stat. 682), as
2 amended, is amended by renumbering title XI (as in effect
3 prior to the enactment of this Act) as title XII, and by re-
4 numbering sections 1101 through 1114 (as in effect prior to
5 the enactment of this Act) and references thereto, as sections
6 1201 and 1214, respectively.

7 (c) The Public Health Service Act is further amended
8 by adding after title X the following new title:

9 "TITLE XI—SICKLE CELL ANEMIA PREVENTION
10 PROGRAM

11 "GRANTS FOR SICKLE CELL SCREENING AND COUNSELING
12 PROGRAMS

13 "SEC. 1101. (a) The Secretary is authorized to make
14 grants to and enter into contracts with public and nonprofit
15 private entities to assist in the establishment and operation
16 of voluntary sickle cell anemia screening and counseling
17 programs and to assist in developing and making available
18 information and educational materials relating to sickle cell
19 anemia to all persons requesting such information or mate-
20 rials, and to inform the public generally about the nature of
21 sickle cell anemia and the sickle cell trait.

22 "(b) In making grants and contracts under this section
23 the Secretary shall take into account the number of persons
24 to be served, the extent to which such screening and coun-
25 seling is needed on a local basis, the relative need of the

1 applicant, and its capacity to make rapid and effective use
2 of such assistance.

3 “(c) For the purpose of making payments pursuant to
4 grants and contracts under this section, there are authorized
5 to be appropriated \$25,000,000 for the fiscal year ending
6 June 30, 1973; \$25,000,000 for the fiscal year ending
7 June 30, 1974; and \$25,000,000 for the fiscal year ending
8 June 30, 1975.

9 “DEMONSTRATION GRANTS

10 “SEC. 1102. (a) In order to promote research in the
11 diagnosis, treatment, and prevention of sickle cell anemia,
12 development of programs to educate the public regarding the
13 nature and inheritance of the sickle cell trait and sickle cell
14 anemia, and the development of centers for research, testing,
15 counseling, prevention, or treatment of sickle cell anemia the
16 Secretary is authorized to make grants to public or non-
17 profit private entities and to enter into contracts with public
18 or private entities and individuals for projects for research
19 and research training in such fields.

20 “(b) For the purpose of making payments pursuant to
21 grants and contracts under this section there are authorized
22 to be appropriated \$5,000,000^{1/} for the fiscal year ending
23 June 30, 1973; \$5,000,000^{1/} for the fiscal year ending June
24 30, 1974; and \$5,000,000^{1/} for the fiscal year ending June
25 30, 1975.

^{1/} In H.R. 11421, introduced by Mr. Anderson, this figure is \$15,000,000.

1 "VOLUNTARY PARTICIPATION

2 "SEC. 1103. The participation by any individual in any
3 program or portion thereof under this title (whether by
4 grant or contract) shall be wholly voluntary and shall not be
5 a prerequisite to eligibility for or receipt of any other service
6 or assistance from or to participation in, any other program.

7 "APPLICATIONS

8 "SEC. 1104. A grant under this title may be made upon
9 application to the Secretary at such time, in such manner,
10 containing and accompanied by such information as the Sec-
11 retary deems necessary. Each application shall—

12 "(1) provide that the programs and activities for
13 which assistance under this title is sought will be admin-
14 istered by or under the supervision of the applicant;

15 "(2) describe with particularity the programs and
16 activities for which assistance is sought;

17 "(3) provide for strict confidentiality of all test
18 results, medical records, and other information regarding
19 screening, counseling, or treatment of any person
20 treated, except for (A) such information as the patient
21 (or his guardian) consents to be released; or (B) statis-
22 tical data compiled without reference to the identity of
23 any such patient;

24 "(4) provide for appropriate consultation with

1 community representatives in the development and op-
2 eration of any program funded under this title;

3 “(5) set forth such fiscal control and fund account-
4 ing procedures as may be necessary to assure proper dis-
5 bursement of and accounting for Federal funds paid to
6 the applicant under this title; and

7 “(6) provide for making such reports in such form
8 and containing such information as the Secretary may
9 reasonably require.

10 “PUBLIC HEALTH SERVICE FACILITIES

11 “SEC. 1105. The Secretary shall establish a program
12 within the Public Health Service to provide for voluntary
13 sickle cell anemia screening, counseling, and treatment.
14 Such program shall be made available through facilities of
15 the Public Health Service to any eligible person requesting
16 screening, counseling, or treatment, and shall include notifi-
17 cation of all eligible persons of the availability and voluntary
18 nature of such programs.

19 “REPORTS

20 “SEC. 1106. (a) The Secretary shall prepare and sub-
21 mit to the President for transmittal to the Congress on or
22 before April 1 of each year a comprehensive report on the
23 administration of this title.

24 “(b) The report required by this section shall contain
25 such recommendations for additional legislation as the Secre-
26 tary deems necessary.”

1 PROTECTION OF ARMED FORCES PERSONNEL

2 SEC. 4. (a) The Secretary of Defense is authorized and
3 directed to promulgate rules and regulations to provide for
4 screening and counseling of members of the Armed Forces
5 (including their dependents), civilian employees of the De-
6 partment of Defense, and persons examined at Armed Forces
7 examining and entrance stations, for the sickle cell trait and
8 sickle cell anemia.

9 (b) Such rules and regulations shall provide for—

10 (1) voluntary screening for the sickle cell trait for
11 persons described in subsection (a) who request such a
12 test, at no cost to such person;

13 (2) communication to such person described in
14 subsection (a) of the results of such test;

15 (3) voluntary referral of individuals determined to
16 possess a positive trait to an appropriate military or
17 civilian counseling or treatment agency;

18 (4) notification to persons described in subsection
19 (a) of the cost-free and voluntary nature of the screen-
20 ing and referral programs implemented pursuant to this
21 section;

22 (5) education of persons described in subsection
23 (a) regarding the nature and inheritance of the sickle
24 cell trait and sickle cell anemia; and

25 (6) assurance that all information obtained on
26 specimens submitted voluntarily under this Act shall be

1 held confidential except for (A) such information as
2 the patient (or his guardian) consents to be released
3 or (B) statistical data compiled without reference to
4 the identity of any such patient.

5 (c) The Secretary of Defense shall provide for volun-
6 tary counseling or treatment of such persons described in
7 subsection (a) found to have the sickle cell trait or sickle
8 cell anemia at an appropriate military or civilian facility as
9 the case may be.

10 (d) (1) The Secretary of Defense shall prepare and
11 submit to the President for transmittal to Congress on or
12 before April 1 of each year a comprehensive report on the
13 administration of this section.

14 (2) The report required by this subsection shall con-
15 tain such recommendations for additional legislation as the
16 Secretary of Defense deems necessary.

17 (e) The participation by any individual in any pro-
18 gram or portion thereof under this section shall be wholly
19 voluntary and shall not be a prerequisite to eligibility for
20 or receipt of any other service or assistance from, or to par-
21 ticipation in, any other program.

22 PROTECTION OF VETERANS

23 SEC. 5. (a) Chapter 17 of title 38, United States Code,
24 is amended by adding at the end thereof the following new
25 subchapter:

3 "§ 651. Notification and education

9 “(b) The Administrator shall establish a program of
10 education regarding the nature and inheritance of sickle cell
11 trait and sickle cell anemia and make such program avail-
12 able to all such eligible persons.

14 “(a) The Administrator shall furnish to any person
15 eligible for care under this chapter who makes a request
16 screening for sickle cell trait or sickle cell anemia.

17 “(b) Upon a finding that such eligible person has the
18 sickle cell trait or sickle cell anemia the Administrator shall
19 provide for voluntary counseling or treatment as the case
20 may be.

22 “(a) The Administrator shall prepare and submit to the
23 President for transmittal to the Congress on or before April 1
24 of each year a comprehensive report on the administration of
25 this subchapter.

1 “(b) The report required by this section shall contain
2 such recommendations for additional legislation as the Ad-
3 ministrator deems necessary.

4 **“§ 654. Voluntary participation**

5 “The participation by any individual in any program or
6 portion thereof under this section shall be wholly voluntary
7 and shall not be a prerequisite to eligibility for or receipt of
8 any other service or assistance from, or to participation in,
9 any other program.”

10 (b) The analysis at the beginning of such chapter is
11 amended by adding at the end thereof:

 “SUBCHAPTER VI—SICKLE CELL ANEMIA PREVENTION

 “651. Notification and education.

 “652. Screening and treatment.

 “653. Reports.

 “654. Voluntary participation.”

DEPARTMENT OF DEFENSE,
DEPARTMENT OF THE ARMY,
Washington, D.C., July 20, 1971.

HON. HARLEY O. STAGGERS,
Chairman, Committee on Interstate and Foreign Commerce, House of Representatives, Washington, D.C.

DEAR MR. CHAIRMAN: Reference is made to your request to the Secretary of Defense for the views of the Department of Defense on H.R. 7654, 92d Congress, a bill "To amend the Public Health Service Act to provide for the establishment of a National Sickle Cell Anemia Institute." The Department of the Army has been assigned responsibility for expressing the views of the Department of Defense on this bill.

The purpose of the bill is to establish within the Public Health Service an institute to be known as the National Sickle Cell Anemia Institute to conduct and support research, provide training, determine instances of the disease and conduct and support educational programs and counseling concerning the disease through the aforesaid Institute. The Institute would also assist the Secretary of Defense in screening all persons entering the armed forces for sickle cell anemia and its traits.

Inasmuch as enactment of the bill would not affect the operations of the Department of Defense, the Department of the Army on behalf of the Department of Defense defers to the views of the Department of Health, Education, and Welfare as the agency having primary interest in this matter.

The enactment of this bill will cause no apparent increase in budgetary requirements of the Department of Defense.

This report has been coordinated within the Department of Defense in accordance with procedures prescribed by the Secretary of Defense.

The Office of Management and Budget advises that from the standpoint of the Administration's program, there is no objection to the presentation of this report for the consideration of the Committee.

Sincerely,

ROBERT F. FROEHLKE,
Secretary of the Army.

DEPARTMENT OF DEFENSE,
DEPARTMENT OF THE ARMY,
Washington, D.C., January 6, 1972.

HON. HARLEY O. STAGGERS,
Chairman, Committee on Interstate and Foreign Commerce, House of Representatives, Washington, D.C.

DEAR MR. CHAIRMAN: Reference is made to your request to the Secretary of Defense for the views of the Department of Defense on H.R. 11171 and H.R. 11742, 92d Congress, bills "To provide for the prevention of sickle cell anemia." The Department of the Army has been assigned responsibility for expressing the views of the Department of Defense on these bills.

The purpose of the bills is to establish a national program for the prevention and treatment of sickle cell anemia by grants, contracts, and through Public Health Service facilities. In addition, the bills authorize and direct the Secretary of Defense to promulgate rules and regulations to provide for voluntary screening and counseling or treatment of members of the Armed Forces (including their dependents), civilian employees of the Department of Defense, and persons examined at Armed Forces Examining and Entrance Stations, for sickle cell trait and sickle cell anemia. The bills also provide for similar treatment and counseling programs by the Veterans Administration.

The Department of the Army on behalf of the Department of Defense defers to the Department of Health, Education, and Welfare as to the desirability of Sections 1, 2, and 3 of the bills and we defer to the Administrator of the Veterans' Administration as to the merits of Section 5. The Department of the Army on behalf of the Department of Defense, however, does not favor Section 4 of the bills for the reasons stated below.

Section 4 provides for a comprehensive program for dealing with the sickle cell trait and sickle cell anemia that would include education, screening, counseling and treatment. The benefits of this program would be available to members of the Armed Forces and their dependents, civilian employees of the Department of

Defense, and persons examined at Armed Forces Examining and Entrance Stations. We are opposed to the inclusion of the latter two categories of personnel in the military health programs of the Department of Defense.

Sickle cell disease is disqualifying under the standards of medical fitness for appointment, enlistment, and induction in the Armed Forces. Most persons with the disease have extensive medical histories to document unfitness for military service. Therefore, it is unnecessary to screen for the disease and few individuals, if any, who have the disease are inducted.

Sickle cell trait is not, in general, disqualifying for military service. Individuals who have the sickle cell trait may, if otherwise qualified for military service, enter active duty and can be cared for under existing military health programs. Consequently, the only persons examined at the Armed Forces Examining and Entrance Stations to whom benefits would accrue under the bills would be those who were identified as having the disease or the trait but who did not enter on active duty. Thus, their entitlement to military health benefits would turn on the fact that they were examined at a facility operated by military personnel. We believe any education, identification, counseling, or treatment for such persons should be under programs available to the general public.

Department of the Army civilian employees are, in general, entitled to the same benefits, including health benefits, as individuals employed by other Federal agencies. While the Department of Defense would support any program designed to aid Federal employees, we believe any such program should not be restricted to employees of the Department of Defense. We defer to the Civil Service Commission as to the desirability of a program for sickle cell anemia and as to the scope of such a program. We would object, however, to a program that relief upon the Department of Defense military health programs. The Department of Defense and Department of Health, Education, and Welfare are, at the present time, conducting a joint study required by Public Law 92-129 to devise means of reducing reliance on health personnel of the Armed Forces in fulfilling our medical needs. Accordingly, we believe it would be undesirable to add civilian employees as additional beneficiaries to the military health programs based solely upon a particular disease or trait of an individual.

If Section 4 of the bills were amended to exclude persons who are examined but do not enter active duty and to exclude civilian employees, then the benefits of Section 4 would be restricted to members of the Armed Forces and their dependents. We believe that the military departments already have sufficient legal authority for effective programs to deal with the sickle cell trait and disease. In fact, many of the objectives of Section 4 are being accomplished under existing authority. For example, we have encountered some problems among service members in association with high altitude, hot and dry climate, and strenuous exercise. Therefore, service members under consideration for Special Forces, flight or diving training are screened for the trait and are ineligible for those duties if the trait is present. It should be noted that, with respect to such duties, we have not permitted a desire to identify the sickle cell trait only on a voluntary basis to overshadow the need to protect individuals through mandatory screening of training applicants.

In view of existing military health programs and our capability to integrate new scientific knowledge into the military health system, the Department of the Army on behalf of the Department of Defense believes that additional legislation specifically covering sickle cell anemia among members of the Armed Forces and their dependents is unnecessary at this time.

The fiscal effects of this legislation are not known to the Department of Defense.

This report has been coordinated within the Department of Defense in accordance with procedures prescribed by the Secretary of Defense.

The Office of Management and Budget advises that from the standpoint of the Administration's program, there is no objection to the presentation of this report for the consideration of the Committee.

Sincerely,

ROBERT F. FROEHLKE,
Secretary of the Army.

DEPARTMENT OF HEALTH, EDUCATION, AND WELFARE,
Washington, D.C., August 26, 1971.

HON. HARLEY O. STAGGERS,
*Chairman, Committee on Interstate and Foreign Commerce,
House of Representatives,
Washington, D.C.*

DEAR MR. CHAIRMAN: This letter is in response to your request of May 3, 1971, for a report on H.R. 7654, a bill "To amend the Public Health Service Act to provide for the establishment of a National Sickle Cell Anemia Institute."

The bill would provide for the establishment of a National Sickle Cell Anemia Institute, with an Advisory Council to the Secretary of Health, Education, and Welfare, and for the initiation of activities, either intramural or extramural or both, in the following areas as they relate to sickle cell anemia: research and training programs and diagnosis, treatment, and prevention; screening programs to determine incidence among children; and counseling and education programs to increase awareness of the characteristics and problems associated with this disease. The activities of the proposed Institute would also include assistance to the Secretary of Defense in the screening of all susceptible persons for the sickle cell trait, entering the Armed Forces.

In his Health Message of February 18, 1971, the President recognized the problem of sickle cell disease and the FY 1972 budget of the National Heart and Lung Institute identifies \$5 million for this special health initiative. The National Heart and Lung Institute is to be the lead agency for the program which is to be coordinated by Dr. Robert L. Ringler, Deputy Director, NHLI. The operational plan of the sickle cell anemia program which has been accepted by me as Secretary of HEW has four major goals: to evaluate the best mode of therapy for sickle cell crises, to develop new knowledge on the fundamental biology of the disease, to ascertain the feasibility and value of community screening for sickle cell disease and sickle cell trait, and to increase public knowledge and awareness of the disease through education and counseling. A Sickle Cell Anemia Advisory Committee is being established to make recommendations concerning the planning, execution, and evaluation of all aspects of the program. This committee, which will be representative of, and responsive to, the needs of the affected community, will be meeting soon.

With so many of the activities proposed in H.R. 7654 already underway within the present organization of the Department, it would seem unnecessary to establish a new Institute to achieve these ends. Furthermore, the lead agency concept, the appointment of a specific coordinator for the program, and the establishment of an Advisory Committee on sickle cell anemia should provide the focus and visibility for this disease problem comparable to that which a new Institute could provide and do so without the prolonged lead time necessitated by the establishment of a new Institute and without the dispersion of money and duplication of effort which such an Institute would require.

We would therefore recommend against enactment of H.R. 7654.

We are advised by the Office of Management and Budget that there is no objection to the presentation of this report from the standpoint of the Administration's program.

Sincerely,

(S) ELLIOT L. RICHARDSON, *Secretary.*

EXECUTIVE OFFICE OF THE PRESIDENT,
OFFICE OF MANAGEMENT AND BUDGET,
Washington, D.C., August 18, 1971.

HON. HARLEY O. STAGGERS,
Chairman, Committee on Interstate and Foreign Commerce, House of Representatives, Washington, D.C.

DEAR MR. CHAIRMAN: This is in response to your request of May 3, 1971 for the views of this office on H.R. 7654, a bill "to amend the Public Health Service Act to provide for the establishment of a National Sickle Cell Anemia Institute."

In its report to your Committee on H.R. 7654, the Department of Health, Education, and Welfare points out that many of the activities proposed in the bill are already being performed within the Department. Therefore, the creation

of a new National Sickle Cell Anemia Institute would cause delay and lead to a duplication of effort.

We concur in the views expressed in the report of the Department of Health, Education, and Welfare and, accordingly, recommend against enactment of H.R. 7654.

Sincerely,

WILFRED H. ROMMEL,
Assistant Director for Legislative Reference.

VETERANS' ADMINISTRATION,
OFFICE OF THE ADMINISTRATOR OF VETERANS' AFFAIRS,
Washington, D.C., December 27, 1971.

HON. HARLEY O. STAGGERS,
Chairman, Committee on Interstate and Foreign Commerce, House of Representatives, Washington, D.C.

DEAR MR. CHAIRMAN: This will respond to your request for a report by the Veterans Administration on H.R. 11171, 92nd Congress, a bill "To provide for the prevention of sickle cell anemia."

The purpose of the bill is stated in its title. It would seek to accomplish that purpose by adding programs to be carried out by the Public Health Service, the Department of Defense, and the Veterans Administration. My comments on the bill will be restricted to those provisions which would be carried out by the Veterans Administration.

Section 5 of the bill would amend chapter 17 of title 38, United States Code, by adding a new subchapter VI—Sickle Cell Anemia Prevention, under which the Administrator would be required to screen and treat all veterans eligible for VA medical care with regard to sickle cell trait and sickle cell anemia.

At the present time, we are engaged in a cooperative study in about twelve VA hospitals, each with about 1500 admissions of black patients. This study will evaluate the increased risks of complicating diseases associated with sickle cell trait. Conditions being studied include pulmonary embolism, myocardial infarction, vascular complications of diabetes, pyelonephritis, and post-operative complications.

Moreover, in addition we have approximately 35 individual research projects underway to study the genetic defects and pathologic mechanisms involved in sickling. In fiscal year 1972 we have committed research funds of approximately \$350,000 to individual projects, with an additional \$200,000 for the cooperative study.

The new section 651, which would be added to title 38 by the bill, requires the Administrator to notify all persons eligible for care under this chapter of the availability of screening, treatment, and counseling programs with regard to sickle cell trait and sickle cell anemia. The key phrase in that provision—"persons eligible under this chapter"—is not precise and presents difficulties in implementation. Apparently, what is desired is that anyone eligible for inpatient or outpatient care be encompassed within that phrase. Under current law, VA hospital care for a non-service connected condition can be furnished to any war veteran or a veteran of service after January 31, 1955, *if he is unable to defray the expenses of necessary hospital care*, or if he is over the age of 65 years. Therefore, since eligibility for the great number of veterans is premised on "inability to defray expenses of hospital care", it would be impossible for the Veterans Administration to identify, as a preliminary to notifying, all persons as required by this provision of the bill.

The proposed new section 652 would require the Administrator to screen any person eligible for care under chapter 17 of title 38, United States Code, who makes a request for screening for sickle cell trait or sickle cell anemia, and to provide treatment and counseling to those who are found to have such trait or disease. Thus, this bill would, for the first time, authorize the Veterans Administration to provide purely screening examinations for a particular disease, independent of an application for hospital admission, medical services, or for compensation, or pension benefits administered by the Veterans Administration.

Our interest in finding a solution to the problems arising from the sickle cell trait is evident from the research we are already conducting in this field as

noted above. To the extent that inpatient treatment would be needed, in most cases we would require no new authority since the bill limits treatment to persons eligible for medical care under title 38.

To the extent that outpatient treatment is needed, we might point out that legislation (H.R. 10880) to broaden our authority to furnish medical services on an ambulatory or outpatient basis passed the House of Representatives on October 4, 1971. That legislation, when enacted, would enhance our ability to identify and treat sickle cell trait and sickle cell anemia as well as the complicating diseases associated therewith.

In addition to certain technical difficulties with the language of the bill, we believe that there is a serious question as to whether the mission of the Veterans Administration should be enlarged to include screening examinations of individuals *independent* of any need for hospitalization or the providing of medical services. At the present time, there is insufficient medical data to define a course of suitable treatment for individuals who have been identified as having the sickle cell trait. Under these circumstances, we believe the more prudent and medically effective approach is to continue with the intensive research program outlined above, rather than divert VA physicians into an extensive screening program, as proposed in this bill. Moreover, the enactment of this provision requiring screening and examination for sickle cell trait and sickle cell anemia would be a precedent for requests to expand our medical services to screening for other diseases. Thus, we cannot recommend favorable consideration of this section of the bill.

It is not possible to make a precise estimate of the cost to the Veterans Administration should H.R. 11171 be enacted, because of our lack of experience in this area of medical care. There are approximately two million black veterans in our population, with one million of those under the age of 45 years. It is reasonable to assume that approximately one-half of those under 45 would seek the screening examination. Under the conditions prescribed by the bill, we estimate that each examination would cost approximately \$17.50. Therefore, to staff and examine this number would cost a total of \$10.5 million. We would expect to accomplish these examinations over a five year period, after which the costs would reduce to a routine for new veterans. A more detailed estimate for each of the first five years follows:

Fiscal year:	Costs (in millions)
1973 -----	\$2.3
1974 -----	2.3
1975 -----	2.3
1976 -----	2.3
1977 -----	1.3
Total -----	10.5

The views expressed herein with regard to H.R. 11171 would be equally applicable to H.R. 11257, H.R. 11420, H.R. 11791, H.R. 11872, H.R. 11924, H.R. 11742, H.R. 11907, and H.R. 11421, similar purpose legislation.

Advice was received from the Office of Management and Budget with respect to a similar report to the Chairman, Senate Committee on Labor and Public Welfare, on similar purpose bill S. 2676, 92nd Congress, that there was no objection to the presentation of the report from the standpoint of the Administration's program.

Sincerely,

DONALD E. JOHNSON, *Administrator.*

Mr. ROGERS. The committee is pleased to have a number of our colleagues here this morning to testify on this proposed legislation.

Our first witness this morning is our distinguished colleague from the District of Columbia the Honorable Walter E. Fauntroy.

We welcome you to the committee and we will be pleased to receive your testimony at this time.

STATEMENT OF HON. WALTER E. FAUNTROY, A REPRESENTATIVE
IN CONGRESS FROM THE DISTRICT OF COLUMBIA

Mr. FAUNTROY. Thank you, Mr. Chairman and members of the committee.

I appreciate this opportunity to speak on H.R. 11171. I more especially appreciate the fact that this subcommittee has moved as rapidly as it has on this measure. To me, this speed indicates the concern of the Congress about the tragedy of this dreaded disease, sickle cell anemia, which affects primarily black people causing their death and heaping upon them a pain and suffering which no one should have to bear. It is a pain and suffering which is made harder because of the dearth of programs and the dearth of funds which could, if available, perhaps find the cure and an alleviation of the affliction.

Sickle cell anemia strikes one of approximately every 500 young black children, killing more than half of them before they reach the age of 20. It is a genetic malady whose horrors are passed from generation to generation recurring predictably when the necessary genetic conditions are present. The gene is normally passed onto the children of parents who both carry the sickle cell trait. Approximately one in 12 blacks carry the trait with the chance of both parents carrying the trait being one in 144.

Blacks who inherit the disease itself suffer periodic crises in which the normally spherical red blood cells are distorted into crescentlike shape. These deformed cells block narrow capillaries, depriving nearby tissues of needed oxygen and causing excruciating pain. Persons who have only the trait generally don't need care for this condition, as it most often does not cause any difficulty except in instances of extreme stress at certain altitudes or during certain phases of other ailments.

Each major crisis for those having the disease—and there is an average of three per year—requires an average of 7 days' hospitalization. In addition, there are generally four to five less serious crises that, while they do not require hospital treatment, require varying degrees of restricted activity. It is not necessary to detail the distraught feeling and the financial loss that affect parents whose children are suffering with this disease.

Until just recently, however, no one considered sickle cell anemia to be the health problem that it is. As a result, there is no nationwide volunteer organization devoted to sickle cell anemia. There are a variety of local groups, but they are not coordinated to mount a national attack and educate the public as are those groups whose concerns are muscular dystrophy, acute leukemia, and cystic fibrosis.

Yet, the problem instead of decreasing, as are many others, is becoming greater. The majority of blacks now live in urban areas. For these areas, because of the incident rate, the disease becomes not just a significant health problem, it has become a prevalent health problem demanding immediate attention. To understand the meaning of this, one must look at the incident rates for other major congenital and genetic childhood diseases. Muscular dystrophy occurs at rates of one per 5,000 births, cystic fibrosis at one per 2,940, and diabetes mellitus at one per 2,500. While sickle cell anemia occurs at the rate of one per 500, in 1967, there were 1,155 cases of sickle cell anemia, 813 of muscular dystrophy, and 1,206 of cystic fibrosis.

In the District of Columbia, where the black population approaches 70 percent, it is estimated that the costs of care for the 1,000 afflicted children approaches \$4 million. And, the disease far exceeds childhood diabetes, nephrosis, or cystic fibrosis as a cause of illness.

Like many other diseases, the key to effective palliative treatment is early diagnosis. Today, there are inexpensive screening tests which can be offered prior to marriageable age.

Those who are found to carry an abnormal hemoglobin gene could be counseled to be sure that their mates were tested. Only in this way can heterozygote pairs be detected, and only in this way can informed decisions be made about childbearing among "at risk" parents. This is not to suggest that a couple will decide to have no children; it is to suggest that whatever they do will be done on the basis of an informed and intelligent decision.

The bill, H.R. 11171, which is now before you has four central provisions: (1) A coordinated Federal grant program of \$25 million per year for 3 years for research, voluntary screening, and counseling and public education; (2) a demonstration grant program of \$5 million per year for 3 years for the development of centers for research and research training in sickle cell anemia; (3) a requirement that the Department of Defense implement a policy to provide voluntary screening, counseling, treatment, and education concerning the disease for servicemen, civilian employees, and inductees; and (4) similar programs by other Federal agencies which provide direct health care for persons eligible in such agencies; namely, the Veterans' Administration and Public Health Service.

Since the purpose of this bill is to provide a coordinated and overall effort against sickle cell disease, a primary intent of the bill, in addition to research, is the providing of funds for the creation of community screening and counseling programs. I have already outlined the need for this. Another provision provides funds for programs to educate the public about the nature of sickle cell anemia and sickle cell trait. This is particularly important in achieving maximization of the counseling and screening services to be offered. In the October 1970 issue of JAMA, Dr. Robert Scott points out that many people do not know anything about the disease—much less know about its effects.

The second major thrust of the bill provides demonstration funds for the development of centers for research, screening, counseling or treatment. Funds would be available to public and nonprofit private institutions such as medical schools, hospitals and foundations. A principal focus of this provision would be to provide funds for centers such as the very fine one at our own Howard University in the District of Columbia, under the direction of Dr. Roland Scott.

The third thrust of the bill enables the Armed Forces and other Federal agencies to offer screening, counseling, and treatment. This voluntary program is especially important because it is here that large numbers of young people can be reached, treated, and educated. Since there are conditions under which persons with the trait can be adversely affected, screening would enable these persons to be identified enabling them to receive treatment and consideration for certain duties which would be more amenable to this health program.

Finally, other available programs which now exist in the Public Health Service are authorized to screen, counsel, and treat. Hopefully, this will permit the many other persons who are not otherwise directly covered to obtain a determination of possession of the trait, and such treatment or counseling as is desirable.

Again, I wish to thank you for allowing me the opportunity to appear and testify on this bill. I hope that you will rapidly approve it. I am sure you will.

Thank you, Mr. Chairman.

Mr. ROGERS. Thank you very much, Mr. Fauntroy, for an excellent statement. We appreciate your presence here today and your interest in doing something about this disease.

Mr. Symington?

Mr. SYMINGTON. Congressman Fauntroy, this is a very helpful statement for us. I am confident that this committee will take early action on this legislation. I, myself, was appalled at the affliction figures you gave us. This is one of these diseases that I am afraid many of us think are rather minimal in national importance until somebody reminds us that it is not the case.

I certainly want to say I welcome your testimony. I hope that we can do something constructive on this bill.

Thank you.

Mr. FAUNTROY. Thank you, Mr. Symington.

Mr. ROGERS. Congressman Carter?

Mr. CARTER. Thank you, Mr. Chairman.

It is true that sickle cell anemia has not received the recognition it should have throughout our country. I was very fortunate as a medical student to have as my teacher Dr. W. L. Diggs in Memphis, Tenn., who has done as much research as any man in the United States on this disease.

I hope that we can authorize funds for more research and certainly for more planning previous to marriage for the people who are afflicted by this condition. At present that seems to me to be the best solution to prevent the people who carry the genes from mating and producing offspring which will not live more than likely beyond the age of 20, and being afflicted with a very insidious pain which goes with the disease.

I thank the gentleman for his testimony.

Mr. ROGERS. Thank you so much for your presence today.

Mr. FAUNTROY. Thank you, Mr. Chairman.

Mr. ROGERS. Also present today we have a group of Congressmen and Congresswomen who wish to present statements before the committee on this important legislation. In order that we may hear from them all and because of the lack of time this morning I believe it would expedite matters if I just called you in order and if you would please come forward and present your statement, we may proceed on that basis.

Our next witness is the Honorable George P. Miller of California. Welcome, sir.

**STATEMENT OF HON. GEORGE P. MILLER, A REPRESENTATIVE IN
CONGRESS FROM THE STATE OF CALIFORNIA**

Mr. MILLER. Mr. Chairman, I am pleased to have the opportunity to appear before this distinguished subcommittee in behalf of H.R. 8598, and related bills, which propose the creation of a national program for the prevention of sickle cell anemia.

In comparison with the research on other fatal diseases, relatively little has been done to seek a cause and cure for this disease. Sickle cell anemia affects approximately one out of every 500 black children. Out of those affected less than half live beyond 20 years of age.

Funds are needed to stimulate research, provide screening programs, and to conduct counseling and educational programs for those suffering from the disease and the general public.

There is no known cure for sickle cell anemia. The establishment of an institute to study this painful and nearly always fatal disease is past due. I urge the committee to give favorable consideration to this humane legislation.

Mr. ROGERS. Thank you, Mr. Miller, for your concise statement.

Next we shall hear from the Honorable Charles C. Diggs, Jr., of Michigan. Welcome, sir.

**STATEMENT OF HON. CHARLES C. DIGGS, JR., A REPRESENTATIVE
IN CONGRESS FROM THE STATE OF MICHIGAN**

Mr. Diggs. Mr. Chairman, I appreciate the opportunity to present my views on the importance of the health problem you are considering today—the tragic and so often deadly disease called sickle cell anemia. It is a blood disease that affects one out of every 10 black people, either directly or as carriers.

Sickle cell anemia results from the inheritance of a genetic factor relating to the sickle cell trait. People may be carriers of this trait without being seriously handicapped. However, if two people who have sickle cell traits marry, chances are one in four that any child they have will have the disease itself; and less than half of those children who contract the disease survive beyond the age of 20.

To date, the only tool available for fighting this disease is a simple blood test to determine whether a person is a carrier. Effort and support must be mobilized to diagnose, prevent, and cure sickle cell anemia through increased research in the cause and treatment of the disease, and the education, screening and counseling of carriers of the sickle cell trait.

I therefore strongly urge your favorable consideration of legislation authorizing sufficient funds for the operation of such programs and services as are necessary to focus and develop effective national leadership in the first to control and cure this dreadful disease.

Thank you, Mr. Chairman.

Mr. ROGERS. Thank you, Mr. Diggs.

Our next witness is from the State of Wisconsin, the Honorable Henry S. Reuss. Welcome Mr. Reuss. Proceed as you see fit, sir.

**STATEMENT OF HON. HENRY S. REUSS, A REPRESENTATIVE IN
CONGRESS FROM THE STATE OF WISCONSIN**

Mr. REUSS. I appreciate this opportunity to restate my support of the bills being considered by your committee—most specifically H.R. 8598—which calls for establishment of a National Sickle Cell Anemia Institute and development of programs for the prevention of sickle cell anemia.

The disease, afflicting almost exclusively black Americans, has been shamefully neglected in the past, as research, fundraising efforts and public attention focused on other—often less prevalent—diseases.

The spotlight has at least been shifted, in recent months, to this inherited blood disease, and the bills now under consideration give hope that the newly awakened interest can be translated into real progress toward its control and eventual eradication.

Consider these facts and the past lack of attention to sickle cell anemia becomes all but incredible:

The disease is an intensely painful one, whose varied effects can include strokes and seizures, chronic bone infections, enlarged heart and liver and jaundice. It kills over half its victims before the age of 20; few survive beyond the age of 40. Most of those afflicted are severely crippled long before death.

Hereditary in nature, the disease is passed down through a trait carried by between 8 and 13 percent of American Negroes. Research indicates that more than 2 million Americans carry the trait.

The disease itself strikes about 1 of every 500 black persons, from among the children of unions between two carriers. An estimated 50,000 American Negroes have the disease; at least 5,000 of these require hospitalization each year with thousands of others treated on an outpatient basis.

In 1967, there were an estimated 1,155 new cases of sickle cell anemia, compared with 1,206 of cystic fibrosis and 813 of muscular dystrophy—more or less similar levels of all three. Yet, in 1968, only \$50,000 was raised for sickle cell research by voluntary organizations, as compared with \$2 million for cystic fibrosis, and \$7.2 million for muscular dystrophy. This represents a striking disparity. Another measure of research support is the number of grants awarded by the National Institutes of Health. Again for 1968, the figures showed 66 for muscular dystrophy, 65 for cystic fibrosis, and only 22 for sickle cell anemia—less even than the number for such exceedingly rare conditions as glycogen storage diseases.

The bills you have under consideration at these hearings would help to set the balance straight by providing for a coordinated Federal Government effort aimed at prevention and treatment of the disease.

Importantly, the legislation provides for public education to reduce the demonstrated lack of knowledge about this dread disease.

It provides for nationwide screening programs, both among school children and among persons entering the Armed Forces, to find and alert those who carry the trait. At the same time, it sets the stage for concentrated research into the mechanisms of sickle cell anemia, toward eventual eradication of the disease.

Nothing less than the comprehensive programs outlined in these measures can be effective in dealing adequately with this long-neglected problem.

Mr. ROGERS. Thank you, Mr. Reuss.

Our next witness is the Honorable Robert N. C. Nix of Pennsylvania.

**STATEMENT OF HON. ROBERT N. C. NIX, A REPRESENTATIVE IN
CONGRESS FROM THE STATE OF PENNSYLVANIA**

Mr. NIX. Mr. Chairman, I am pleased to have this opportunity to present my statement in behalf of legislation to create within the National Institutes of Health a new Institute devoted to sickle cell anemia.

As you know this disease is especially prevalent among Negroes. It afflicts one out of every 500 black children and causes the death of many of them before the age of 20. For these and for many others it is a very painful, as well as deadly, ailment.

Yet, I regret to say, we have not as a Nation devoted to this disease even a significant fraction of the money we have applied to research into other childhood diseases. Apart from a few isolated studies, very little has been done to determine better ways of detecting and anticipating this hereditary affliction or to devise more effective means of treating the disease once it occurs.

The proposed National Sickle Cell Anemia Institute could give effective national leadership and support to develop more effective means of dealing with this disease. This leadership could result in the development of more adequate programs of diagnosis, treatment and prevention throughout the country. National screening programs are clearly called for, to identify the carriers of the hereditary sickle cell factor and to make available to these carriers the proper counseling to avoid the matching of these deadly genes in future generations.

The training of more medical manpower for research in this area is also clearly indicated and the institute could give leadership in this area, as well as provide more research opportunities for the medical manpower we should train.

We have recognized the value of the National Institute of Health approach since the first of these Institutes, the National Cancer Institute, was established in the 1930's. Many victims of cancer, who would have died through our ignorance and neglect in earlier years, are now being saved as a result of the work of the Cancer Institute. This is also true of heart disease and of many other diseases.

I urge you to make it true also of sickle cell anemia. We cannot excuse the failure to make a national commitment to combat this disease simply because it is a very serious problem only to one part of the population. It is serious to the one person, whatever his race, who is affected. And to my people it is a major health problem. It is one entitled to the attention of the Nation.

Mr. ROGERS. Thank you, Mr. Nix.

Mr. NIX. Thank you, Mr. Chairman.

Mr. ROGERS. Next we shall hear from the Honorable William S. Moorhead. Mr. Moorhead is also from the State of Pennsylvania.

STATEMENT OF HON. WILLIAM S. MOORHEAD, A REPRESENTATIVE
IN CONGRESS FROM THE STATE OF PENNSYLVANIA

Mr. MOORHEAD. Thank you, Mr. Chairman. Sick cell anemia has always been present in the population of black citizens of this Nation. This disease is a part of the Negro's heritage and one which cannot be corrected at this time. Despite the fact that sick cell anemia has been estimated to affect seriously more than 100,000 individuals, it is a relatively unknown disease. It has also been estimated that 8 to 13 percent of American Negroes carry the sick cell trait and thus must give this problem serious consideration whenever marriage and family planning are considered. While this inherited trait apparently provides some protection against malaria, in the United States, where malaria is under control, the trait does not provide even this marginal usefulness and the disease is a serious health problem.

The lack of support given to research and treatment for the prevention and cure of sick cell anemia in the past has been due in part to the absence of any coordination of effort to combat this disease. It is a fact that sick cell disease in the black citizen has not been a high priority public health problem in comparison with other frequently less common diseases. There have been a few dedicated and devoted investigators who have studied this disease and struggled to secure support for their work. It is largely due to their efforts that any progress has been made at all. Much of this work has been performed in the absence of adequate funding and frequently without any Federal support at all. Voluntary groups have not organized on a national basis to secure funds and as a result of this fragmentation of effort progress has been very slow.

There is much work which has to be completed. Sick cell anemia has been ignored for such a long time that public health authorities have not even provided adequate attention to informative education programs to alert and advise blacks of the potential dangers from the disease. A majority of blacks are totally unaware of the facts which are known about sick cell disease.

Tests are being developed for the identification of sick cell factor in citizens who carry the hereditary trait and to assist in the confirmatory diagnosis of those who actually have the disease. Some of these tests are quite good and can be made available for large scale screening. It is a poor promise, however, which identifies a problem without offering further advice and medical assistance. This disease, particularly severe in children, and psychologically frustrating to carriers of the trait, has no really acceptable cure. Physicians can alleviate much of the suffering from the disease when it is diagnosed but no entirely satisfactory treatment is available. And what of the individuals who carry the trait? Once the individual with the trait is identified, expert genetic counseling must be made available, to provide assistance in making family planning decisions.

Of course, the long-range objective is to devise a technique or procedure to eliminate the disease altogether. Whether such a procedure will come from investigations in biochemistry or from more exotic genetic engineering cannot be envisioned at this time. Certainly, the cure will never occur in an acceptable time interval if financial support is not given to the investigator of the future.

The bill which I have proposed, H.R. 11104—a bill to amend the Public Health Service Act to provide for the establishment of a National Sickle Cell Anemia Institute, has several major objectives. One of these objectives is to focus attention on this terrible disease in order to secure a coordination of the Nation's efforts to control the disease. The struggle to treat and prevent this disease has gone on too long in a low-keyed and fragmented fashion. We need to offer the resources of the Federal biomedical research community to investigate this disease and bring new and stronger efforts to bear on the problem. By establishing a National Sickle Cell Institute, biomedical administration and funding programs will receive the necessary direction and guidance to assure full support of all objectives of my bill. Other objectives include—

1. The conduct and support of research on sickle cell anemia;
2. The training of medical manpower for diagnosis, treatment, and prevention of the disease;
3. To provide for the education of the Nation's blacks as to the dangers of the disease and make genetic counseling service available; and
4. To provide screening services for members of the Armed Forces for sickle cell anemia and its traits.

Sickle cell anemia is not a rare and exotic disease. Similar and less frequently encountered diseases found in the Caucasian race have been identified and screening and treatment services provided. Sickle cell anemia occurs with a frequency much higher than other important childhood diseases which have been given much more support. Surely, we have ignored this disease long enough. The adoption of H.R. 11004 would serve notice that we acknowledge our responsibility to confront this need with a high order of priority.

Mr. ROGERS. Thank you, Mr. Moorhead, the committee appreciates your statement.

Mr. MOORHEAD. Thank you, Mr. Chairman.

Mr. ROGERS. The Honorable Joseph P. Addabbo of New York is our next witness. Welcome, sir. Please be seated.

STATEMENT OF HON. JOSEPH P. ADDABBO, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF NEW YORK

Mr. ADDABBO. Mr. Chairman, I welcome this opportunity to present testimony in support of legislation to provide for the establishment of a National Sickle Cell Anemia Institute. The disease known as sickle cell anemia is one which has not received a great deal of publicity until recently. Now that the problem has been made the subject of increasing public and congressional interest the facts about this disease have stirred the humane concerns of millions throughout our Nation.

Sickle cell anemia is an inherited and fatal blood disease which can be transmitted to a child when both parents have a condition which can be determined by a blood test. The disease occurs in one out of every 500 births and claims the lives of half its victims before they reach the age of 20. The disease occurs primarily among black Americans and educational efforts have begun to alert the black communities across the Nation of the great dangers.

As a cosponsor of legislation now being considered by this subcommittee, I urge that we in the Congress seize an opportunity to assist in making major breakthroughs in the treatment and prevention of this disease. Some progress has been made already in the area of treatment, however, research efforts are scattered and uncoordinated. The purpose of my bill and the others pending before you is to place the efforts under a single coordinating body established in the Public Health Service.

My bill would therefore create a National Sickle Cell Anemia Institute to coordinate and support programs for diagnosis, treatment and prevention of this disease. The legislation would authorize traineeships in the Institute and in other medical institutions to pursue these goals.

Mr. Chairman, we have made a serious error by not acting to spur progress in this area in decades gone by. Now there is an opportunity to make real progress by assigning a high priority to the conquest of this disease. Just as this subcommittee has taken the lead in framing a new national effort to cure cancer, so must you again take the lead in organizing a similar fight against sickle cell anemia.

The legislation before you requires a very small investment in terms of the human lives which can be saved by its authorization. I urge you to act favorably on the bills to establish a National Sickle Cell Anemia Institute.

Thank you, Mr. Chairman.

Mr. ROGERS. Thank you, Mr. Addabbo.

Next we shall hear from my fellow Floridian, the Honorable Claude Pepper. Welcome, sir.

STATEMENT OF HON. CLAUDE PEPPER, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF FLORIDA

Mr. PEPPER. Thank you, Mr. Chairman. The prevalence of sickle cell anemia in the black population is serious enough today to represent one of the more important diseases in our Nation. Striking one out of every 500 Negro births, sickle cell anemia affects more people than more publicized childhood diseases such as cystic fibrosis which occurs once in every 1,400 births.

Because the disease is particularly severe in children, it claims the lives of half of its victims before the age of 20. Yet, despite the high frequency and the grave statistics of sickle cell anemia, it is a long ignored and little understood disease. The reason for this lack of awareness of sickle cell anemia has been the absence of a coordinated effort on a national scale to educate the affected portion of our population and to sponsor research as a priority effort.

The present work and research being done is quite limited. Various isolated and independent studies on sickle cell anemia are being conducted, but they are without the adequate funding and consolidation needed to insure an effective program. A national coordinated attack on this disease would accelerate progress now being made, and more importantly, provide a concentrated emphasis on finding the most effective treatment and cure for sickle cell anemia.

We have had to face the problem of disease in this Nation before. Cancer and heart disease were more serious threats to the health of this land a few years ago than they are today. At that time I supported the establishment of the National Cancer Institute and the National Heart Institute. The progress and advancement of these two institutes in combating cancer and heart disease should stand as proof of the merit of nationwide programs against disease. I have proposed, therefore, in H.R. 8598 that a similar nationwide program be launched against the disease of sickle cell anemia through the establishment of a Sickle Cell Anemia Institute.

This Institute would conduct and support programs for the diagnosis, treatment, and prevention of sickle cell anemia. Nationwide screening programs would be instituted to determine the frequency of the disease as well as to identify the carriers of the hereditary sickle cell factor. If these carriers are thus identified, expert genetic counseling may then be made available to provide family planning assistance.

By establishing a National Sickle Cell Anemia Institute, the research programs will receive the necessary direction and guidance in investigating this disease. Medical manpower can be trained through the Institute for the diagnosis, treatment, and prevention of the disease. The Institute would educate the Nation's blacks, the chief victims of the disease, as to the dangers of sickle cell anemia as well as informing the people of the genetic counseling services available to them. The Institute would also provide a screening service for members of the Armed Forces for sickle cell anemia and its symptoms.

The establishment of a Sickle Cell Anemia Institute represents the strongest and most effective organizational weapon we can have against this deadly disease.

Mr. ROGERS. Thank you, Mr. Pepper.

Next we shall hear from our colleague from the State of New York, the Honorable Frank Horton. Please be seated, sir.

STATEMENT OF HON. FRANK HORTON, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF NEW YORK

Mr. HORTON. Mr. Chairman, it has often been said that disease is no respecter of boundaries—social, racial, economic, or geographic. Generally, this is true. Cholera outbreaks in the Middle East, for example, are a threat all over the world because of our modern high speed transportation systems and this disease can be a hazard to all—men, women, and children—black, yellow, white, or red.

Recently, however, we have been made aware of one of the more serious exceptions to the ubiquitous nature of disease: sickle cell anemia. Sickle cell anemia is an exception because it strikes only one section of our population—our black citizens.

Sickle cell anemia is a hereditary disease of the blood. This disease, first described in 1910, did not have its biochemical nature characterized under 1949, about 40 years later. Technically, a differentiation must be made between sickle cell trait and sickle cell disease. The former term refers to those individuals who carry only one inherited factor or gene for sickle cell anemia but do not suffer from the disease;

an individual who inherits an abnormal gene from both parents will contract sickle cell anemia. It is normal with this type of inheritance to find more carriers of the disease than those afflicted.

It has been estimated that more than 2 million of our black citizens carry the trait for sickle cell anemia and that there are now more than 50,000 American Negroes who have sickle cell disease. I should point out, Mr. Chairman, 50,000 is a most conservative figure and your committee will undoubtedly hear estimates ranging as high as 100,000. Within the black community, one child in every 500 births inherits a trait from both parents. Very few of them live past early middle age.

These statistics show that the incidence of sickle cell disease is higher than several other childhood diseases which affect all races, such as muscular dystrophy and mongolism. And yet, when the record is examined, we find that our support of biomedical research on sickle cell anemia has not been given the priority which should be expected if priorities for research were determined by the seriousness and incidence of a disease.

We are all aware of the logic behind our national effort to support research for cancer and heart disease. We are reminded, sometimes very personally, that cancer and heart disease show very little discrimination in the selection of the target organism and that these diseases are the two major killers and cripples in our society. If incidence is one of the criteria for the adoption of priorities in allocating our limited biomedical research resources, it is difficult to understand the relatively low priority which has been given to the study of sickle cell anemia.

Of course, I fully understand that many other factors, such as progress in related sciences, may be involved. But history shows us that sickle cell disease has been known for more than 60 years and the fundamental nature of its biochemical cause for more than 20 years. An examination of the work on this disease reveals that the progress which has been made is the result of the work of a few very devoted and brilliant individuals—in frequent instances, the dogged, determined efforts of black physicians working with limited resources.

Moreover, Mr. Chairman, we have been notably remiss in making available to our black citizens the knowledge which has been developed. We know how to test for the presence of the trait and to confirm the diagnosis of the disease. Yet, there is no routine national program to examine blacks to identify the sickle cell trait. There is no national program to provide genetic counseling services to carriers of the trait in order to assist them in making decisions in their family planning. Clinical evaluation of promising treatments for those suffering from the disease is carried on in a very limited way. In other words, it appears that our Federal medical apparatus has not been used as effectively as it might have been to confront this problem in a coordinated and forthright manner.

Recently, President Nixon indicated that he considered sickle cell disease to be a problem requiring national attention. The level of effort, in Federal health agencies has been increased to about \$6 million, whereas previous funding levels had been near \$1.5 million. Although this increased attention is certainly welcome, it is my opinion that it is too little and quite late. After 60 years of dragging our feet,

we can and should devote more resources to the problem of sickle cell anemia. The real problem, I believe, is not that there is resistance to accelerating our attack on sickle cell disease, but rather with the determination of how much we can provide and how we should go about formulating our battle plan.

A number of proposals have been made to accelerate our progress toward the prevention and cure of sickle cell disease. H.R. 8597, which I sponsored together with 17 of my colleagues, proposes that a National Sickle Cell Anemia Institute be established. Certainly the formation of such an institute would be a strong organizational technique for administering an attack on this disease. We have many precedents for such a procedure. The National Cancer Institute was established with this type of objective in mind—a coordinated attack on a specific group of diseases. The sickle cell disease program within the National Institutes of Health is scattered through several institutes—genetics in one area, blood research in another, and biochemistry in another. From a management standpoint, it seems that the structuring of research and health services within one organization would do much to coordinate our efforts on sickle cell disease.

Mr. Chairman, I am well aware of the intense debate with regard to the administrative structure for expansion of our efforts on cancer research. I am also familiar with the arguments against setting up separate institutes of research for each of the various diseases of mankind. The fact remains that we must find a way to conduct our program for diagnosis, treatment, and prevention of sickle cell disease more effectively. We need to strengthen considerably our research programs on this disease; we need to provide for the training of personnel for the diagnosis, treatment, and prevention of sickle cell anemia; we need to provide for nationwide screening to determine the incidence of the disease, to educate our black citizens to the hazards of the disease, and to provide counseling services. Finally, we owe it to the black members of our Armed Forces to provide these same services.

I support H.R. 8597 to establish a National Sickle Cell Anemia Institute as one way of accomplishing these objectives. I hope in our consideration of our responsibilities to the black citizens of this Nation that we move quickly and effectively to satisfy the objectives of this bill. If there are serious objections to the establishment of a separate institute, then an alternative proposal offering essentially the same program should be agreed upon. In any event, Mr. Chairman, we should not allow ourselves to become so deeply embroiled in the administrative aspects of the program that our progress toward the solution of the problem is slowed or seriously reduced in scope.

Mr. ROGERS. Thank you, Mr. Horton.

Next we shall hear from Hon. William J. Green of Pennsylvania.

STATEMENT OF HON. WILLIAM J. GREEN, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF PENNSYLVANIA

Mr. GREEN. Mr. Chairman and subcommittee members, I thank you for the opportunity to offer you my reasons for supporting legislation to provide funds for research and treatment of sickle anemia cell.

Sickle cell anemia is a national problem. I believe, however, that because it is a disease which afflicts black people exclusively, that its urgency has virtually gone unnoticed.

This disregard for sickle cell anemia is manifested in a number of unfortunate ways.

The amount of money spent on sickle cell research is meager. What is worse, there is no reliable treatment for the thousands of blacks presently suffering from the disease.

I am certain, though, that other witnesses have adequately provided the facts about sickle cell anemia, so I will not repeat these tragic details. I will, however, fill you in on the efforts being made in my own city, Philadelphia, to control sickle cell. For one, October was declared "Sickle Cell Month" in Philadelphia * * * a month to draw attention to the problem which could afflict the children of 20,000 blacks who live in the city, because they carry genetic trait for the disease.

Philadelphia is also the home base of VASCA, Volunteers in Aid of Sickle Cell Anemia, which was organized there in 1959, as the Nation's first lay organization to focus attention on the problem. Last month, VASCA, led by its president, Mrs. Julia Davis, began a fundraising drive to aid victims of sickle cell anemia.

In the areas of treatment and research, two Philadelphia physicians have performed notable work. Dr. Frank Gardner, director of hematology at the Presbyterian University of Pennsylvania Medical Center, has developed a new method of blood transfusion—replacing sickled cells with new blood cells. But the treatment is only temporary, for transfusions cannot go on forever.

Dr. Christopher Holroyd, an instructor at Pennsylvania Medical School, has developed an urea in sugar solution which has had some success in unsickling red blood cells.

There has also been a limited program established to test pupils for sickle cell anemia in some of Philadelphia's junior high schools.

I can only hope that these efforts will help focus attention on the problem and stimulate further efforts toward a cure and eventual elimination of sickle cell anemia.

I am concerned about the almost total neglect that the problem has received throughout the Nation and the limitations imposed by a lack of funds on those projects, such as the ones I have mentioned in Philadelphia, which are working for a solution to the problem. Therefore, I strongly urge the committee to give serious consideration and favorable action to the bill before you.

The bill provides the funds sorely needed for research, counseling, public education and voluntary screening, in the civilian and government sectors. Further, it provides for a demonstration grant program for the development of centers for research and training in sickle cell anemia.

We must move now to pass this legislation. We can no longer ignore a disease that afflicts more than 100,000 black Americans.

Mr. ROGERS. Thank you, Mr. Green.

Mr. GREEN. Thank you, Mr. Chairman.

Mr. ROGERS. Our colleague from the State of New York, the Honorable Frank J. Brasco, is our next witness.

Come forward, sir. Please have a seat and proceed as you see fit.

**STATEMENT OF HON. FRANK J. BRASCO, A REPRESENTATIVE IN
CONGRESS FROM THE STATE OF NEW YORK**

Mr. BRASCO. Mr. Chairman, I very much appreciate this opportunity to testify before you today on behalf of a measure I have joined in sponsoring. I refer, of course, to H.R. 8597, which would amend the Public Health Service Act to provide for establishment of a National Sickle Cell Anemia Institute.

In recent months, significant facts have come to public light regarding this ailment. Black Americans are the victims more often than not. Usually, it strikes at children, afflicting them in such a painful manner that none of its victims are able to live beyond the middle thirties.

Scientific findings published in recent months indicate that there is a very real chance to do something about this disease, if a concentrated effort is made. A national commitment has been made in other areas, and is required here. If the Nation decides to do something about this ongoing situation, we could conceivably curb its ravages.

Unfortunately, the state of the scientific art in relation to this affliction has a long way to go in order to make significant inroads into its present position. For this reason, I have joined in sponsoring this bill, which would set up a special institute to deal with the problem.

It would be charged with developing and supporting programs for diagnosis, treatment and prevention of sickle cell anemia. A concerted, national effort would be launched under the aegis of the Public Health Service to do something about this program.

The Fellowship of Black Athletes has already testified on behalf of such an endeavor. They have come up with facts that cannot be refuted. Surely, we here in the Congress can do what we can to hold up our end of this effort.

It is one of the shames of this Nation that large minorities among us are afflicted with illness that erodes the quality of their lives. Some, as in the case of sickle cell anemia, are always fatal.

We beat our breasts in public about this state of affairs, times without number, yet little is actually done about it. Such a situation is known to the groups of people so affected. Certainly black Americans are aware of and protesting this state of affairs.

The Congress has the power to act in this case. Delay means more deaths of innocents, and by inaction in the future, we share some of the blame for them. Hopefully, we shall take this opportunity to act now.

Mr. ROGERS. Thank you, Mr. Brasco.

Our next witness is our colleague from the full committee, the Honorable Dan Kuykendall of Tennessee.

Welcome, Mr. Kuykendall.

**STATEMENT OF HON. DAN KUYKENDALL, A REPRESENTATIVE IN
CONGRESS FROM THE STATE OF TENNESSEE**

Mr. KUYKENDALL. Chairman Rogers, gentlemen of the committee, it is a pleasure to appear before you this morning to express my wholehearted support for H.R. 11171, the National Sickle Cell Anemia Prevention Act.

As you all know, the funds that would be authorized by this bill are divided into two parts—\$25 million a year for the next 3 fiscal years for screening and counseling, and an additional \$5 million a year for basic research and testing of treatment methods. In other words, sociological funds and scientific funds.

That is precisely the way I view the entire problem of sickle cell anemia—as a two-headed monster, sociological and medical.

I think the distinguished junior Senator from California and I are in complete agreement, that the crying need in this field, and the general goal of this legislation, should be to increase public awareness and public understanding of sickle cell anemia.

It is undoubtedly true that heart disease and cancer kill more black Americans each year than does sickle cell anemia, but most Americans, black and white, are well aware of the danger signals associated with these dread diseases.

Few Americans, black or white, have ever heard of sickle cell anemia, let alone appreciate its danger signals and complications.

I wonder how many of the Congressmen serving on this subcommittee who feel they know something about this disease realize that in many cases it requires emergency treatment.

Sickle cell anemia is excruciatingly painful. A terrorizing collection of symptoms attend this disease. But there is more to it than pain and suffering.

The sociological problems of sickle cell anemia must be considered along with what we hope will be an ultimate scientific cure.

We have found in our attempts in demonstration projects in the city of Memphis, which has one of the largest concentrations of black people in the Nation, that if this problem is not properly presented to the people, there appears immediately a stigma which makes the citizenry reluctant to submit to a screening test.

We have found that the best way to explain this disease is to give the true historical background. Being a carrier of the sickle cell anemia trait is not a weakness. It is not a stigma. Actually, it is a historic strength. The sickle cell trait is historically a protection from malaria. It developed through the centuries among residents of malarial areas.

Progress is being made at the University of Tennessee medical units in Memphis in one area of sickle cell research—that of chemotherapy. It is for this area of research that I have been trying to get additional funds for the University of Tennessee. Using chemotherapy doctors in Memphis have been able to reduce the length of attacks and their severity to some extent.

One of the byproducts, and I think it is a completely natural one among the young, due to the intensive pain of sickle cell anemia, is the tendency to turn to narcotics. Sickle cell anemia victims turn to drugs not for the reasons that other young persons do, but simply to get relief from the excruciating pain. So in short we must not only educate those people who might be carriers of the sickle cell trait, but we must also educate the rest of the country to the seriousness and the full ramifications of the disease.

The genetic charts that you use in studying sickle cell anemia are very clear. If two carriers marry each other, half of their children will

have the disease itself and will be doomed to a life of pain. However, if carriers do not marry each other, there is practically no possibility of an infected child emerging.

Congressman Fauntroy ably pointed out recently that it is a more prevalent disease than leukemia, than cystic fibrosis, and actually more painful.

That is how I got interested in the subject—and I must give credit here to two of the outstanding researchers in the field, a husband and wife team at the University of Tennessee in Memphis who have spent a great many years in this field. They are Dr. Lorraine Kraus, who is a Ph. D. and Dr. Alfred Kraus, a distinguished M.D.

My personal involvement has been sharpened and intensified by my association with these two experts, but it stems from something much more basic than that—it stems from the simple fact that there are a hell of a lot of people in my District affected by sickle cell anemia.

The Krauses tell me, and I think it is generally accepted, that between 8 and 10 percent of the Nation's black population has the sickle cell trait, and that one out of every 40 of those with the trait actually suffers from sickle cell anemia. Translate that into my district—just one congressional district in this Nation.

There are approximately 720,000 people in Shelby County, Tenn. Of those 36.9 percent, or about 266,000 are black. Using the Krauses percentages, that means from 22,000 to almost 27,000 people are walking around with the trait of this disease.

If we had 22,000 people in Memphis with a cystic fibrosis trait—just the trait, mind you, not the disease itself—we'd have a public health crisis that would make headlines all over the world.

We must do something, we must do much, much more than we are doing, to attack sickle cell anemia on every front—through continuing research, and above all, through education of the people, education of those susceptible to it to recognize its symptoms, education of our medical professions to treat it, education of the rest of us—and that includes the Congress—to give sympathetic understanding and help where it is needed.

A measure of that help and understanding is now being given by an organization that deserves much credit—the Black Athletes Foundation. Led by Willie Stargell, the cleanup hitter of the world champion Pittsburgh Pirates, and Muhammad Ali, the former heavyweight champion, this private foundation is doing much to alert the public to the symptoms and danger signs, and to urge blacks to take a simple blood test.

One of the most popular areas of public misunderstanding—and here is where we must take immediate steps toward our educational efforts—is that the trait is not the disease. The person carrying the trait is exactly like the man whose second cousin may be mentally retarded. He himself is perfectly healthy, but if he were to carry someone whose grandmother had a retarded child, that couple might possibly have a retarded child themselves. That is a trait, just like blue eyes are a trait, just like sickle cell blood is a trait.

Nobody, particularly if he or she looks healthy and feels healthy, likes to be told he has a disease. The word carrier brings to all of us the image of "Typhoid Mary." A large dose of education, seasoned

with a milligram of salesmanship, could go a long way toward removing the stigma that the very word "disease" carries with it. I strongly suggest that the words "disease" and "carrier" be stricken from our vocabularies when we are talking about the trait.

I was heartened, just yesterday, to learn that the National Heart and Lung Institute has allocated almost half of its funds for this year for education and counseling. As you all know, President Nixon took the initiative earlier this year, earmarking \$5 million of the Nation's health budget for sickle cell research and counseling, and the Heart and Lung Institute is now geared to putting its money where it will do the most good.

Mr. Chairman and gentlemen of the committee, now is the time for Congress to single out sickle cell anemia as a national objective, and to give our researchers and our sociologists the money they must have to conquer that objective.

Mr. ROGERS. Thank you, Mr. Kuykendall, for a fine statement.

Mr. KUYKENDALL. Thank you, Mr. Chairman, for affording me the opportunity to be here today.

Mr. ROGERS. Next we shall hear from the Honorable Glenn M. Anderson of California.

Welcome, sir. Please be seated.

STATEMENT OF HON. GLENN M. ANDERSON, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF CALIFORNIA

Mr. ANDERSON. Mr. Chairman, sickle cell anemia kills half of its victims before their 20th year, and most of the rest by age 40.

This painful and deadly blood disease is inherited when both parents carry the sickle cell trait. Approximately 2,500,000 black Americans carry this trait and pass it along to their offspring. Approximately one black child out of every 400 to 500 born in the United States inherits this often fatal disease.

What is the solution? How can we attack this killer?

There are techniques for examining individuals to detect whether the sickle cell trait is present. The most common technique is the Sickledex test which costs about 50 cents per blood sample to perform. However, for those who have the disease no completely satisfactory cure is presently available. The reason for the lack of information on sickle cell anemia is inadequate funding.

Simply to fund a Sickledex test for the residents of the city of Compton, Calif., would cost about \$27,890.

The National Sickle Cell Anemia Prevention Act, which I introduced, would create a preventive program in the National Institutes of Health.

To assist the communities in the establishment and operations of voluntary sickle cell anemia screening and counseling programs, the Secretary of Health, Education, and Welfare would be authorized to spend \$75 million over the next 3 years.

This legislation also authorizes \$45 million over the next 3 years to promote research in the diagnosis, treatment, and prevention of sickle cell anemia.

In order to provide detection, counseling, and treatment for black veterans and members of the armed services, the Administrator of the Veterans' Administration and the Secretary of Defense are directed to establish voluntary programs.

Mr. Chairman, we must direct our immediate attention to the problem of sickle cell anemia, and we must make every effort to cure this severe health problem. An effective national program, such as envisioned in this legislation, would produce a significant benefit to the black community, specifically, and our entire country, generally.

Mr. ROGERS. Thank you, Mr. Anderson.

Our next witness is from our full committee. Welcome, Mr. Metcalfe.

**STATEMENT OF HON. RALPH H. METCALFE, A REPRESENTATIVE
IN CONGRESS FROM THE STATE OF ILLINOIS**

Mr. METCALFE. Mr. Chairman, a few years ago, scientists discovered the cause of a disease that has been afflicting millions of individuals of black ancestry since the beginning of time.

Until now, however, very little attention was given to this disease, and often people with this affliction went for a lifetime without ever knowing that such a disease existed, not to mention that they themselves had the disease and that it might be transferred to their offsprings.

At present, an estimated 50,000 black Americans have sickle cell disease. At least 5,000 require hospitalization each year and many others are treated as outpatients in hospital emergency rooms. In addition, these patients are subject to general lassitude due to anemia, chronic infections, and other complications. Most never live beyond the age of 30.

The most important and effective measure of decreasing incidents of sickle cell anemia is prevention through what is called "genetic counseling." This means that the black population must become aware of the disease and of its inherited trait, and that blood tests can and should be given to detect the 8-plus percentage of Americans who are carriers of this trait.

This Congress should establish programs for the treatment and control of this disease that affects such a large number of black Americans.

I have cosponsored three bills which will provide funds for the detection, treatment, and research of sickle cell anemia.

I cosponsored a bill which will provide for the establishment of a National Sickle Cell Anemia Institute. The Institute will conduct and support comprehensive programs for the diagnosis, treatment, and prevention of sickle cell anemia; establish related traineeships in the Institute and elsewhere; establish nationwide screening programs to determine incidents and traits among school age children; develop counseling and education programs in consultation with community representatives to make known the services available under this legislation; and provide assistance to the Secretary of Defense for screening persons entering the Armed Forces for the disease.

I also cosponsored a supplemental appropriations bill that would restore the \$6 million that was cut from the Health Education, and

Welfare appropriations for 1971-72 to aid current programs for the detection, treatment, and research of the disease.

In addition to these two bills, I have cosponsored a sickle cell anemia proposal to attack the sickle cell problem on a local level. This bill would provide funds for local community efforts to screen for the disease and would make large grants for major research in a pilot program here in the District of Columbia.

In December, the congressional black caucus, along with the National Dental Association, the National Medical Association, Howard University, and Meharry Medical College, will sponsor a "National Health Conference on the Status of Health in the black Community" at Meharry in Nashville, Tenn.

The conference will be divided into eight discussion workshops, one of which will be a section on special minority problems. Sickle cell anemia will be discussed in this workshop and hopefully, participants in this conference will bring this disease, along with other social health problems affecting the black community, to an appropriate level of concern for the total black population.

If sickle cell crises can be avoided through treatment and prevention, much greater control of this disease may be possible in the future. Only extensive clinical trials and better understanding of the disease can enable those afflicted to live longer with less pain and disability.

I know that the members of this Subcommittee on Health and Environment, as well as all the members of the Interstate and Foreign Commerce Committee, are as deeply concerned as I am about this problem. And, I believe that we all are hopeful that the most effective, comprehensive, nationwide program will be developed legislatively for the diagnosis, treatment, and prevention of sickle cell anemia, as a result of this subcommittee's dedicated consideration under the very able leadership of its Chairman Paul Rogers.

Thank you, Mr. Chairman.

Mr. ROGERS. Thank you, Mr. Metcalfe.

Our next witness is the Honorable Ella T. Grasso of Connecticut. Welcome, Mrs. Grasso.

STATEMENT OF HON. ELLA T. GRASSO, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF CONNECTICUT

Mrs. GRASSO. Mr. Chairman, there is no greater tragedy than that of a youngster who from birth is unable to fulfill the bright promise of his life.

Yet, for some children, an inherited and deadly blood disease known as sickle cell anemia places severe restrictions on his ability to live a full and active life.

I heartily commend you and your subcommittee. For you are focusing attention on the need to develop an effective treatment and cure of this disease by holding hearings on this important legislation. Sickle cell anemia has long been ignored—a neglect which has become a national disgrace.

In May of this year, 31 of my colleagues cosponsored my bill to create a National Sickle Cell Anemia Institute. The legislation—H.R. 7654, H.R. 8597, and H.R. 8598—which your subcommittee is consid-

ering today, provides for the establishment of a coordinated national attack on this grave threat to many of our people. The National Sickle Cell Anemia Institute, would conduct and support programs for the diagnosis, treatment and prevention of sickle cell anemia. The bill would set up traineeships in the Institute and elsewhere for matters relating to the diagnosis, treatment and prevention of sickle cell anemia.

This legislation would initiate nationwide screening programs to determine the incidence of sickle cell anemia and its traits among school-age children. Counseling and education programs would be developed in consultation with community representatives in order to make individuals and communities aware of the services available under this legislation.

Since an excellent opportunity exists to attend to the health needs of members of our Armed Forces, this bill provides assistance to the Secretary of Defense for screening persons entering the Armed Forces for sickle cell trait.

In addition, to oversee the work of the Institute, an advisory board would be established to consult with and make recommendations on matters relating to the Institute.

Mr. Chairman, it is indeed heartening to know that so many Members of Congress are concerned that proper attention be given to the treatment and cure of this long neglected disease. Until recently, very little effort has been made by the Government to conduct research on sickle cell anemia, which is more prevalent than many of the more highly publicized childhood diseases. Last year, funds in excess of \$1 million were spent by the National Institutes of Health for research of sickle cell anemia. The National Institute of Arthritis and Metabolic Diseases granted over \$600,000 for the study of the disease. This year, preliminary plans of the Department of Health, Education, and Welfare advisory committee on sickle cell disease call for the distribution of \$6 million of recently allocated funds for a program of research and community service. According to the advisory committee chairman, the emphasis of the program will be on "the solution to the problems emanating from sickle cell disease control and eventual eradication."

In March the Foundation for Research and Education in Sickle Cell Disease announced that a nationwide drive will begin to alert black Americans to the peril of sickle cell anemia. Financed through a \$50,000 grant from the Chase Manhattan Bank Foundation, the program will consist of a nationwide network of volunteer groups to make potential carriers aware of the disease and what they can do about it.

My own State of Connecticut has been a leader in bringing to the public's attention the need for priority status to find a cure for sickle cell disease. Through the dedicated and constructive efforts of Mr. Leonard J. Patricelli, president of WTIC-TV, broadcast house in Hartford, a series of editorials and special programs informed viewers of sickle cell anemia and its effects. Mr. Patricelli's efforts were a catalyst for a fund drive in the Greater Hartford community which netted some \$30,000 for the benefit of the Center for the Study of Sickle Cell Anemia at Howard University. School nurses in Hartford

are offering health counseling to children who are found to have sickle cell trait, and other programs on a more permanent basis are being established.

Throughout Connecticut and in my own Sixth District, citizens have responded to the need to help victims of sickle cell disease. Last spring the Concern organization of northwest Connecticut sponsored a dance and raised over \$700 which was sent to Howard University for its sickle cell anemia research program. In August, several citizen action groups held a sickle cell testing day in Torrington for the benefit of area residents.

These and many other commendable activities in this area are certainly encouraging. They have raised the level of national consciousness about this terrible disease. Also, progress is being made in early detection of the sickle cell trait among children, as well as in new procedures for treating the disease. However, unlike proposed campaigns to conquer the threat of cancer and other diseases, there exists no coordinated, properly funded national attack on the grave problem of finding the most effective treatment and cure for sickle cell anemia. Such a national effort would greatly accelerate the progress now being made to lessen the agony caused by this disease.

It is for this reason that I have introduced H.R. 7654, H.R. 8597, and H.R. 8598. Over 2 million Americans carry the trait of this genetic blood disorder—but because they are not ill, they are unaware that they can transmit this trait or the illness to their children. Mr. Chairman, we must direct our immediate attention to the problem of sickle cell anemia, and we must make every effort to cure this severe health problem.

I strongly urge the subcommittee to give favorable consideration to H.R. 7654, H.R. 8597, and H.R. 8598. The Congress will then have the opportunity to assert its proper leadership in generating a much needed national effort to ease the frightful burdens now endured by thousands of people afflicted with this cruel disease.

Mr. ROGERS. Thank you, Mrs. Grasso, for a fine statement.

Mrs. GRASSO. Thank you, Mr. Chairman.

Mr. ROGERS. Our next witness is Mrs. Hicks of Massachusetts. Welcome, Mrs. Hicks. Please be seated and proceed as you see fit.

STATEMENT OF HON. LOUISE DAY HICKS, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF MASSACHUSETTS

Mrs. HICKS. Mr. Chairman and members of the committee, thank you for permitting me to appear before you in support of H.R. 8423, a bill to establish a National Sickle Cell Institute, and H.R. 10935, a bill making a supplemental appropriation for the Secretary of Health, Education, and Welfare for detection and treatment of, and research on, sickle cell anemia.

My district which is the Ninth Congressional District in Massachusetts lies within the city of Boston.

According to the U.S. Census, 1970, as reported by the Research Bureau Boston Redevelopment Authority: In Boston 104,483 of the 641,000 citizens are of black ancestry; this equals approximately 16.3 percent. The census of 1970 almost certainly underestimated the actual population of black citizens and there has been a definite increase in

the number of blacks in Boston during the past year or so. It would be realistic to state that the population of blacks in Boston currently numbers between 110,000 and 120,000 which is approximately 20 percent of the population and live in the South End, North Dorchester, and Roxbury areas.

A number of surveys have been conducted in the last 35 to 40 years as to the prevalence of sickle cell trait. The several surveys conducted of urban population give a prevalence level as stated of sickle cell trait of 8.5 percent which means that Boston contains, in its population, approximately 9,000 individuals with sickle cell trait. Accordingly, the prevalence in Boston of sickle cell anemia must be about 180 individuals.

Today I would like to confirm my continuing support for a significant increase in Federal funds for an attack on sickle cell anemia. I introduced, in May of this year, H.R. 8423 in the belief that it was time to establish a priority effort to gain control of and eliminate this terrible disease of black America.

Sickle cell anemia was first identified more than 60 years ago. Research on a cure for this disease was initiated shortly thereafter but the first major breakthrough in demonstrating the molecular basis for the disease did not occur until 1949 when Dr. Linus Pauling, a well-known biochemist and Nobel Prize winner, showed that the characteristic sickling of the red blood cell was due to an abnormal hemoglobin molecule. Just recently, and using Dr. Pauling's work as a basis for the investigation, a small step has been taken toward the evaluation of a new chemical treatment for patients in the acute stages of the disease. We need to exploit this progress which has been achieved if we are to avoid another half-century of awaiting a cure.

This disease is unusual in its occurrence. Sickle cell anemia is limited primarily to black citizens of our country who have inherited the trait for the disease from each of their parents. The evidence suggests that the disease became established in the Negro population as a result of some poorly understood evolutionary processes associated with a resistance to malaria. In Africa, where this disease had its origin, the biochemical protective mechanism afforded blacks with the sickling trait a greater degree of resistance against the lethal effects of malaria than blacks without the trait. As a result this mutation of the hemoglobin molecule has survived.

Individuals who inherit the trait for the disease from each of their parents suffer terribly from the disease. The manifestations of the disease in small children is particularly pitiful to see and patients with the disease seldom live beyond the age of 40. One difficulty in combating the disease has been that the disease requires special testing for identification and diagnosis and this testing has not been readily available.

Federal support for investigation of sickle cell anemia is scattered throughout several institutes of the National Institutes of Health, including the National Institute of Arthritis and Metabolic Diseases; the National Heart and Lung Institute; and the National Institute of General Medical Sciences; and the Health Services and Mental Health Administration. Although coordination of effort has been assigned to the National Heart and Lung Institute, the history of research on this disease indicates that it has never received any priority of attention.

In 1971, President Nixon announced that Federal funds to combat sickle cell disease would be increased to \$6 million. While this level of funding represents almost a fourfold increase in effort, it is not nearly enough to take immediate advantage of the knowledge which is already available and to extend this knowledge to the black population. Clearly what is needed is some means of improving the lot of individuals effected with sickle cell anemia, hopefully by so modifying this aberration of hemoglobin that it does not cause symptoms, or at worse, causes very minor symptoms. This means research. As I have indicated, there have been a few promising signs for the treatment of the disease. Genetic counseling services and screening to identify the presence of the disease could be provided. These services should be provided to their fullest extent.

It is my opinion that one of the major obstacles to a successful confrontation of the problem of controlling this disease is the decentralization of effort which exists now. The attack on this disease requires a strong and centrally controlled administration of effort. The National Sickle Cell Anemia Institute which I am proposing would provide for this direction of effort, and I trust its main thrust would be the underwriting and support of extramural research grants.

There is some indication that the several voluntary organizations which have been trying to secure visibility for the disease and to educate people to its hazards will be coordinating their efforts through affiliation with a newly established National Sickle Cell Disease Research Foundation. Federal efforts in research, diagnosis, screening, genetics counseling, and treatment would profit immeasurably from a similar centralization of effort. The establishment of a National Sickle Cell Anemia Institute would demonstrate also that there is congressional concern with the fact that the disease has been neglected for too long and would assure the black citizens of our country of our intention to provide the continued support necessary to gain control of this disease.

Mr. ROGERS. Thank you, Mrs. Hicks.

Our next witness this morning is the Honorable Charles B. Rangel of New York.

Welcome, Mr. Rangel. Please come forward and proceed as you see fit, sir.

STATEMENT OF HON. CHARLES B. RANGEL, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF NEW YORK

Mr. RANGEL. Mr. Chairman, I appreciate this opportunity to present my statement of the Subcommittee on Public Health and Environment in support of legislation which would commit the Federal Government to an all-out fight against deadly sickle cell anemia.

Unfortunately, it is common for the Congress to belatedly acknowledge the problems of the poor and minority groups in the United States. Look how much time was required to enact lead-based paint poisoning prevention legislation or to fund bilingual education programs. Earlier this month, the House of Representatives killed the ethnic heritage studies provision of the Higher Education Act.

That is why these hearings are such a promising step. We in the black community have waited a long time for congressional action. We watch one out of every 500 black babies born in the United States die from sickle cell anemia by the age 20. We see 10 percent of the black population in this country carrying the sickle cell trait genetically with inadequate means of detection of the trait.

In human terms, what we have experienced can best be described by the graphic tragedy of one patient which appeared in the October 1971 issue of Ramparts magazine.

She is twenty-four years old and has been in the hospital eighteen times. She would get tired easily ever since she was a child; she did not develop as rapidly or as well as the other children; she would often get colds and more respiratory infections. At age seven her tonsils were removed. At age eight she was hospitalized for a month with severe joint pain, fever, and heart murmurs, diagnosed incorrectly as rheumatic fever. At twelve she developed ulcers on both ankles which would not heal, even now, twelve years later. She then began having periodic attacks of severe, incapacitating pain in her bones and joints, her back and her abdomen. These "crises" would last about a week, leaving her exhausted, weaker than ever, and sore all over.

When she was fourteen her appendix was removed. At sixteen her spleen was removed. In the last eight years she has had six more hospital admissions for the treatment of her recurrent leg ulcers which has eaten down to the layer of muscle and bone. Two admissions for pneumonia, two for long episodes of fever, chills, night-sweats and diarrhea. Now, she tells you, she is urinating blood and her belly is swelling like a balloon and her heart, her doctors have told her, is too big and is getting tired.

She cannot sleep lying down, but only on three pillows, and she wakes up at night gasping for breath. Her eyes have been yellow for six years and her liver, she knows is not working right. In her life she has received (if she remembers right) eighty-six transfusions of whole blood.

Research has been painfully slow. Dedicated physicians such as Dr. Roland B. Scott and Dr. Angela Ferguson of Howard University have labored for 20 years, hampered by a lack of critically needed funds.

We have neglected this killing disease for too long. That is why this legislation before you is so important to millions of Americans, many of whom have already given up any hope of substantial Federal action. The slashing of \$6 million earmarked by the Senate to fight sickle cell anemia by the House-Senate conference on the HEW appropriations this past summer is a perfect example of our misguided priorities. We callously refuse to fund sickle cell anemia detection, treatment, and research yet we enact the military procurement appropriations with scarcely a dissenting vote.

H.R. 10932, my bill to establish a National Sickle Cell Anemia Institute, and H.R. 11171, Congressman Fauntroy's bill of which I am a cosponsor to establish a sickle cell anemia prevention program, are now before the subcommittee. Each attempts to deal directly with the problems of sickle cell anemia, rather than leaving programs up to the discretion and good will of the Secretary of Health, Education, and Welfare.

The National Sickle Cell Anemia Institute would conduct and support research programs for the diagnosis, treatment, and prevention of sickle cell anemia and would provide training and instruction in related matters. In addition, it would provide a nationwide screening program for school age children and set up, with adequate funding, community counseling and education services. An advisory council would be appointed by the Secretary of Health, Education, and Wel-

fare to advise, consult with, and make recommendations to him on matters relating to the institute. Finally, the institute would assist the Secretary of Defense in screening GI's for sickle cell anemia and its traits.

H.R. 11171, the National Sickle Cell Anemia Prevention Act, would empower the Secretary of HEW to make grants to and contracts with public and nonprofit private organizations to set up and operate voluntary sickle cell anemia screening and counseling programs and to develop and make available educational materials relating to this dreadful disease. He may also fund demonstration programs. Public Health Service facilities would be made available for screening, counseling, and treatment services. Further, the Secretary would prepare an annual report to Congress on the implementation of this legislation. The Secretary of Defense would be required to promulgate rules and regulations for the voluntary counseling and screening of servicemen and civilian Defense Department personnel, as well as potential enlistees and draftees at Armed Forces examining and entrance stations; he, too, would annually report to Congress and make recommendations for additional legislation.

A critical section of H.R. 11171 is the mandate to the Veterans' Administration to notify veterans of the availability of voluntary screening, treatment, and counseling programs and to furnish care to eligible veterans.

I cannot stress strongly enough the vital nature of the military and veterans provisions in these two bills. Today, a young man with braces on his teeth is exempt from the military draft, but a young man with sickle cell anemia is still considered suited for military duty. What this means, in down-to-earth terms, is that young draftees and enlistees continue to die shortly after going onto active duty. The New England Journal of Medicine on May 14, 1970, recounted the heart-rending deaths of four young men in basic training. The Army had made no attempt to determine whether or not they had sickle cell anemia, and this criminal negligence resulted in their death. It is time for the Congress to tell the Defense Department that our prime concern must be the health of the individual serviceman, that all necessary steps to preserve that health must be taken, and that no American should be forced into military service if that service poses a tangible and lethal danger to his health.

Mr. Chairman, I do not view these two bills as contradictory. Rather they supplement one another. Therefore, it is my recommendation that the subcommittee give serious consideration to combining the two bills. Specifically, I urge you to establish a National Sickle Cell Institute within the National Institutes of Health to assure black Americans of a congressional commitment to defeat this disease. The Institute should be endowed with the powers enumerated in H.R. 10932 and H.R. 11171, with special emphasis on treatment and screening of schoolchildren, GI's, and veterans. It should be funded annually with a minimum of the \$30 million authorized in H.R. 11171. The Institute should assist the Secretary of Defense and the Veterans' Administrator in establishing their own voluntary programs, providing technical and professional assistance where necessary. Annual reports to Congress should be required since it is imperative that Congress be fully aware of the progress and difficulties of the sickle cell anemia programs.

Congress must make a pledge now that it will wage war against sickle cell anemia until every citizen of this country can live free of fear that he, or his children, or his friends, will be stricken by the disease. We must guarantee all Americans the best medical care available. Favorable action by the Subcommittee on Public Health and Environment is crucial if we are to make rapid progress in our struggle against sickle cell anemia.

Mr. ROGERS. Thank you, Mr. Rangel.

Mr. RANGEL. Thank you, Mr. Chairman.

Mr. ROGERS. Our colleague from the State of Wisconsin, the Honorable Les Aspin, is our last congressional witness this morning.

STATEMENT OF HON. LES ASPIN, A REPRESENTATIVE IN CONGRESS FROM THE STATE OF WISCONSIN

Mr. ASPIN. Mr. Chairman, thank you for permitting me to address the subcommittee today on this vital legislation.

The National Sickle Cell Anemia Prevention Act would provide a long overdue remedy to one of our Nation's most serious yet little-known diseases. For far too long we have ignored this debilitating and widespread problem, and it is high time that we launch an attack on sickle cell.

Mr. Chairman, this bill is most ambitious in scope, and comprehensive in nature. It provides for diagnosis, treatment, and prevention of sickle cell anemia, and for voluntary sickle cell anemia screening, counseling, and treatment. In addition, it authorizes the Secretary of Defense to screen all military personnel and persons examined at Armed Forces examining and entrance stations for sickle cell trait and sickle cell anemia.

This last section is of particular concern to me. I have long felt that we should examine all military recruits to see if they carry sickle cell trait. So far, the Department of the Army has refused to conduct full-scale examinations for sickle cell trait, restricting testing to those candidates for certain special types of duties. I hope that we will now extend this testing to all persons examined by the Armed Forces so that we may identify sickle cell trait in all men and women as a matter of course.

Another section of this bill provides for the Veterans' Administration to notify those under their charge of the programs available under this act. This, too, deserves our most serious consideration. We certainly owe our veterans an obligation to give them counseling and treatment for sickle cell, and I would hope that upon passage of this legislation, many veterans will take advantage of this provision.

Mr. Chairman, it is my hope that the subcommittee will see fit to approve this important bill so that we may finally put the Federal Government firmly on record as committed to an all-out attack on this dread disease.

Mr. ROGERS. Thank you, Mr. Aspin, for a fine statement.

Mr. ASPIN. Thank you, Mr. Chairman, for affording me the opportunity to be here today.

Mr. ROGERS. Our next witness is the Deputy Assistant Secretary for Legislation (Health), Dr. John S. Zapp, Department of Health, Education, and Welfare.

Dr. Zapp, we welcome you again before the committee. We will be pleased to receive your testimony.

STATEMENT OF DR. JOHN S. ZAPP, DEPUTY ASSISTANT SECRETARY FOR LEGISLATION (HEALTH), DEPARTMENT OF HEALTH, EDUCATION, AND WELFARE; ACCOMPANIED BY DR. FRANK BECKLES, DIRECTOR, NATIONAL CENTER FOR FAMILY PLANNING SERVICES; AND DR. ROBERT RINGLER, DEPUTY DIRECTOR, NATIONAL HEART AND LUNG INSTITUTE, NATIONAL INSTITUTES OF HEALTH

Dr. ZAPP. Thank you very much, Mr. Chairman, for the opportunity to appear again before your subcommittee.

I have accompanying me this morning on my right Dr. Frank Beckles, Director of the National Center for Family Planning Services.

Mr. ROGERS. Dr. Beckles, we welcome you.

Dr. BECKLES. Thank you, Mr. Chairman.

Dr. ZAPP. On my left is Dr. Robert Ringler, Deputy Director of the National Heart and Lung Institute.

Mr. ROGERS. We welcome you, too, sir.

Dr. ZAPP. Mr. Chairman and members of the subcommittee, I am pleased to appear before you today to discuss the problem of sickle cell disease and more specifically H.R. 11171, the National Sickle Cell Anemia Prevention Act, and related bills, and H.R. 7654 establishing a National Sickle Cell Anemia Institute and related bills. I will use this occasion, also to provide a status report on the sickle cell disease program that has been launched within the Department at the direction of the President.

Before discussing the bill, I would like to describe briefly the disease.

SICKLE CELL DISEASE

Sickle cell disease is an inherited blood disorder. It is due to a genetically determined change in the chemical substance, hemoglobin, which is responsible for the oxygen-carrying capacity of the blood. The presence of this abnormal hemoglobin, commonly referred to as hemoglobin S, leads to distortion of the normally spherical red blood cells. The distorted red blood cells take on the shape of a sickle, hence the name sickle cell anemia.

The disease may occur in two forms: A relatively benign form referred to as sickle cell trait; and the severe form, sickle cell anemia.

Carriers of sickle cell trait typically do not experience ill effects from the disease except under conditions of extreme physical stress or diminished oxygen supply. However, each offspring from two carriers of the trait has a 25-percent chance of having the overt disease, sickle cell anemia, and a 50-percent chance of carrying the trait, and a 25-percent chance of being free of the disease. Said another way, on the average, 1 in 4 of the children would be expected to have normal hemoglobin, 1 in 4 would be expected to have the anemia, and 2 in 4 to have the trait. Since carriers of the trait typically do not experience ill effects from the disease, they may not be aware of the hazard of bearing children with the overt disease if they unknowingly happen to choose a mate who is also a carrier of the trait. It is for this reason, among others, that so much emphasis is placed on screening for the carriers of sickle cell trait and on medical counselling of those found to be positive.

Unlike the carriers of sickle cell trait, patients with sickle cell anemia experience frequent painful crises which are caused by blockage of normal blood flow in small blood vessels by the sickled red cells. Normally spherical red cells easily flow through small blood vessels. The distorted, sickle-shaped red cells tend to form "log jams" which stop or impede the flow, thus depriving the surrounding tissue of normal oxygen supply which in turn causes severe pain in the affected area.

At the present time, treatment of these most painful crises is aimed mainly at relief of pain, although newer forms of therapy offer some promise of our being able to shorten the crisis by hastening reversal of the sickling phenomenon. It must be emphasized that such therapy must still be regarded as being in the investigation phase of development.

Although the so-called sickle cell crises are the most predominant symptom of the disease, the patients also have other symptoms. Their growth and development may be retarded, and they may have leg ulcers, jaundice, kidney problems, and an increased susceptibility to infection in general and pneumonia in particular. Some of these symptoms come from the severe anemia that results from the fact that the sickled cells are easily destroyed and cannot be replaced as fast as they are lost.

Detailed statistics on the prevalence of sickle cell anemia are currently not available. However, it is estimated that about 2 million black Americans are carriers of the trait. A few other ethnic groups of Mediterranean origins have a greatly lower incidence rate. About 1 in 500 black infants is born with sickle cell anemia. This means that approximately 1,000 black infants are born each year with sickle cell anemia and it is estimated that between 25,000 and 50,000 individuals are currently afflicted with it. Sickle cell anemia kills many of its victims before the age of 20, few survive beyond the age of 40 and most are disabled before death. Because sickle cell disease is caused by a genetic abnormality, there is no way known at present to cure the disease in these individuals. As mentioned earlier, current methods of treatment are aimed primarily at alleviating the pain.

BACKGROUND

Sickle cell anemia has been recognized since 1910 when J. B. Herrick first noted the peculiar "sickle" shape of the red blood cells of a West Indian student in Chicago. It was later learned that under conditions of deoxygenation, red blood cells of sufferers from the disease characteristically assume the sickle form. In 1949, Linus Pauling and his coworkers demonstrated that the disease results from the presence of abnormal hemoglobin (hemoglobin S) within the red blood cells of the patient. Many others have contributed to our knowledge of hemoglobin S, including Dr. Makio Murayama, a member of the staff of the National Institution of Arthritis and Metabolic Diseases, who worked out the detailed special configuration of this molecule in his laboratories at NIH. He is continuing his fundamental physiochemical studies of sickling.

The present state of research in sickle cell disease presents a frustrating picture. The molecular basis for the disease is quite well understood from the work of many investigators. Yet researchers have

been unable to produce a form of therapy which prevents the clinical manifestations of sickle cell anemia. Recently, new forms of treatment such as the use of urea have been introduced. These may make it possible to reverse sickling and thereby shorten or even prevent the painful crises.

DESCRIPTION OF H.R. 11171 AND H.R. 7654

Now I would like to turn to the bills before this subcommittee. H.R. 11171 would amend the Public Health Service Act to establish a sickle cell anemia prevention program consisting of two major parts:

1. Authority for grants to and contracts with public and nonprofit private organizations to assist in the establishment and operation of voluntary sickle cell anemia screening and counseling programs and to assist in developing and making available to the public, information and educational materials relating to sickle cell anemia.

2. Authority for grants and contracts for research and research training in the diagnosis, treatment, and prevention of sickle cell anemia; development of programs to educate the public regarding the nature and inheritance of the sickle cell trait and sickle cell anemia; and the development of centers for research, testing, counseling, prevention, or treatment of this disease.

Other provisions of the bill call for a program to be established for eligible persons treated by the Public Health Service; annual reports by the Secretary, HEW, on the administration of the sickle cell anemia program; authorization for the Secretary of Defense to provide for screening and counseling of members of the Armed Forces and their dependents, civilian employees of DOD, and all persons examined by the Armed Forces, for the sickle cell trait and sickle cell anemia; and requiring notification and a program to be set up by the Veterans' Administration for all persons eligible for medical care under title 38, United States Code, for screening, treatment, and counseling with regard to sickle cell trait and sickle cell anemia.

H.R. 7654 would provide for the establishment of a National Sickle Cell Anemia Institute, with an Advisory Council to the Secretary of Health, Education, and Welfare, and for the initiation of activities, either intramural or extramural or both, in the following areas as they relate to sickle cell anemia: research and training program; diagnosis, treatment, and prevention programs; screening programs to determine incidence among children; and counseling and education programs to increase awareness of the characteristics and problems associated with this disease. The activities of the proposed institute would also include assistance to the Secretary of Defense in the screening of all susceptible persons entering the Armed Forces for the sickle cell trait.

ADMINISTRATION INITIATIVES

The seriousness of this disease and the lack of attention paid to it in the past were recognized by the President in his Health Message of February 18, 1971, when he said, "It is a sad and shameful fact that the causes of this disease have been largely neglected throughout our history. We cannot rewrite this record of neglect but we can reverse it." To this end, he proposed increasing the budget for research and treatment of sickle cell disease fivefold, to a new total of \$6 million.

This administration is fully committed to doing everything possible to cope with the many and varied problems of sickle cell disease, and I would like to briefly sketch activities currently underway and planned.

In response to the President's initiative, the Secretary has assigned to the National Heart and Lung Institute, NIH, responsibility for coordinating the joint HEW effort and Dr. Robert L. Ringler, Deputy Director, NHLI, who is here with me today, was designated coordinator for the program.

The NHLI appropriation contains the fiscal year 1972 funds earmarked for this special program. However, it is planned that some of the funds will be made available to other agencies to carry out some segments of the program.

In addition, the Secretary appointed a Sickle Cell Disease Advisory Committee on August 4, 1971, for the purpose of recommending objectives and plans to carry out the President's initiative.

I would like permission to insert in the record the appointments and the names of the people on the Advisory Committee.

Mr. ROGERS. Without objection it is so ordered. (See p. 64.)

Dr. ZAPP. The Committee held its first meeting on August 13 and a second meeting on October 7-8. As a result of these activities, an operational plan for the Sickle Cell Disease program has been developed and is now in the early phases of implementation. The program has four major goals:

(1) To develop and evaluate new modes of therapy for sickle cell crises;

(2) To develop new knowledge on the clinical complications of the disease;

(3) To develop improved methodology for community screening and counseling programs; and

(4) To increase public knowledge and awareness of the disease through education and counseling.

In accordance with the recommendations of the Advisory Committee—and again I would ask consent to have the full recommendations inserted in the record.

Mr. ROGERS. Without objection it is so ordered. (See p. 65.)

Dr. ZAPP. We expect to implement the program in fiscal year 1972 in four major program segments:

(1) \$2.5 million will be allocated for Comprehensive Centers of Research and Service in Sickle Cell Disease, no more than five in number, to be organized around ongoing programs in sickle cell disease and to encompass all aspects of the program, from fundamental research to demonstration community service activities, and including the training of professional and allied health professions personnel.

(2) \$1 million will be allocated for Screening and Education Clinics, 10-20 in number, to develop methodology and serve as models of the community service organizations capable of providing education, screening, definitive diagnosis, counseling, and referral for appropriate follow-up. It is planned that HSMHA will have primary responsibility for this part of the program.

Dr. Beckles, who is accompanying me today, has the responsibility of coordination within HSMHA.

(3) \$1.5 million will be allocated for research aimed at developing improved treatments for sickle cell crises and new knowledge about the clinical consequences of the disease.

(4) A clearing house for educational and informational materials will also be developed. This will provide a focal point for all types of information on sickle cell disease.

The Committee also recommended that the Department of Defense screen all susceptible recruits for sickle cell disease and that the Veteran's Administration carry out a screening and education program.

ONGOING RESEARCH

In addition to these new initiatives, there has been continuing activity within the Department with regard to sickle cell disease.

Ongoing research relevant to sickle cell disease is now being supported by several of the institutes of the National Institutes of Health. As part of its overall effort, the National Heart and Lung Institute supports research in diseases of the blood, both acquired and of genetic origin. Utilizing funds which were already available, and unrelated to the new initiative, the Blood Resources Branch of the NHLI has awarded approximately \$500,000 in grants for the support of the first phase of a cooperative study to test the feasibility of clinical trials of therapy for the sickle-cell vaso-occlusive crisis. While major emphasis is on evaluation of urea therapy, other therapeutic regimens are included. The institute also supports a number of grants with major emphasis on sickle cell disease, at a total current funding level of approximately \$450,000.

The National Institute of Arthritis and Metabolic Diseases is also quite active in the support of sickle cell disease research. I have already mentioned the importance of the fundamental work by Dr. Murayama in his laboratory at NIH. NIAMD research grant support of sickle cell disease studies totals approximately \$420,000. Both NHLI and NIAMD support a number of projects in which the work is related to sickle cell disease. The National Institute of General Medical Sciences and the National Institute for Child Health and Human Development also support projects directed at sickle cell disease, but on a much more limited scale.

SERVICE ACTIVITIES

Within the Health Services and Mental Health Administration, the National Center for Family Planning Services recently established a Sickle Cell Disease Branch. This organization is responsible for coordinating intensified efforts to improve and expand mechanisms in which services are aimed at ameliorating the debilitating effects of sickle cell disease. The scope of these efforts comprise education, definitive diagnosis and counseling. Components of these services are found in the following regional programs:

Use of NCFPS grantee facilities in four regions—New York, N.Y.; Atlanta, Ga.; New Orleans, La.; and Los Angeles, Calif.

Use of Community Health Services resources in four regions—Miami, Fla.; Chattanooga, Tenn.; St. Louis, Mo.; and Denver, Colo.

In addition, screening, diagnosis, and counseling services for sickle cell disease are provided by a large number of Maternal and Child Health Services funded under title V of the Social Security Act.

At the present time, the National Institute of Mental Health has made an initial commitment of approximately \$25,000 to the sickle cell disease effort. This sum represents their initial commitment and should not be construed as placing a ceiling on their effort. It is being used for studies, and improved mechanisms for dealing with psychological impacts of sickle cell disease on the individual, the family, and the community.

SUMMARY

Mr. Chairman, we are supporting the objectives of H.R. 11171 and H.R. 7654, and related bills, now. As we mentioned earlier, the President has recognized the scope of the problem and has targeted an additional \$5 million in fiscal year 1972 for the sickle cell disease program. This is over and above the \$1 million which is currently being spent in research on this disease. Since the activities proposed in H.R. 11171 are already underway within the present organization of the Department or will be implemented in the current fiscal year, we feel it unnecessary to establish new authorizing legislation for the sickle cell disease program. The lead agency concept, the appointment of a specific coordinator for the program, and the establishment of the Sickle Cell Disease Advisory Committee should provide the focus and visibility for this disease problem comparable to that proposed in these bills.

The primary purpose of the program outlined above, at least in its initial phases, is to develop appropriate methodology before mass community efforts are undertaken. The Sickle Cell Disease Advisory Committee stressed the need for such an approach. The committee also cautioned against allowing publicity regarding sickle cell disease and/or screening programs to exceed the capacity for public education and followup for carriers of the trait as well as for patients.

Creation of a new institute to conduct and support research and services in sickle cell disease is an inappropriate response to this serious health problem. Currently, as has already been stated, the sickle cell program involves coordination of the major research efforts of the NIH and the major service efforts of HSMHA, and within the NIH, three institutes are deeply involved in sickle cell research; namely, NIAMD, NIGMS, and NHLI, while other institutes are making contributions to sickle cell research in similar ways on a smaller scale.

To attempt to reorganize all of these facets of the existing program into a single institute for sickle cell disease would involve an extensive and costly effort which would not be likely to promote progress, since substantial human and financial resources would be absorbed in administrative requirements not now necessary and would cut off this program from the many supportive programs now readily available.

Multiplication of institutes will not, of itself, give assurance of progress in solving the scientific problems associated with this disease nor will it give assurance of the generation of an appropriate therapy and technique of prevention.

Proliferation of institutes within the NIH would inevitably raise administrative costs and to complicate and fragment the delicate network of communication that currently exists. The lead agency concept presently being used offers greater promise of achievement in both research and health care delivery for sickle cell disease.

CONCLUSION

The administration has recognized that sickle cell disease has afflicted black people in America for many years. Moreover, only recently has there been widespread awareness of the dimensions of the problem. We believe that we have adequate authority and that the steps I have outlined constitute a solid program for developing an effort to deal with sickle cell disease. We feel that the authority proposed in these bills is redundant and therefore unnecessary. We cannot comment on those provisions of H.R. 11171 which concern the Department of Defense and the Veterans' Administration. We have been working closely with those two agencies, and this is reflected in their representation on the Sickle Cell Disease Advisory Committee through ex-officio memberships.

(Attachments to Dr. Zapp's statement follow:)

SICKLE CELL DISEASE ADVISORY COMMITTEE

- Mrs. Ruth L. Aikens (Chairman), Associate Director for Health, National Urban League, Inc., 55 East 52d Street, New York, New York, 10022.
- Mrs. Arnita Y. Boswell, Field Work Associate Professor, School of Social Service Administration, University of Chicago, Chicago, Illinois 60637.
- Dr. George J. Brewer, Professor of Human Genetics, Department of Human Genetics, University of Michigan, School of Medicine, Ann Arbor, Michigan 48104.
- Dr. William H. Bullock, Associate Professor of Medicine, School of Medicine, Howard University, Washington, D.C. 20001.
- Dr. C. Lockard Conley, Professor of Medicine, Hematology Division, Johns Hopkins Hospital, Baltimore, Maryland 21205.
- Dr. Yvette Fay Francis, 115-77 Farmers Boulevard, St. Albans, New York 11412.
- Dr. Rudolph E. Jackson, Assistant Member in Hematology and Oncology, St. Jude Children's Research Hospital, 332 North Lauderdale, P.O. Box 318, Memphis, Tennessee 38101.
- Mr. Moses J. Newson, Executive Editor, Afro-American Newspapers, 628 North Eutaw Street, Baltimore, Maryland 21201.
- Mr. William B. Robertson, Special Assistant to the Governor, State Capitol, Richmond, Virginia 23219.
- Dr. Louis W. Sullivan, Associate Professor of Medicine, Boston University School of Medicine, Boston, Massachusetts 02118.
- Mrs. Zeola Walder, South Central Multipurpose Health Services Center, 2051 East 103d Street, Los Angeles, California 90002.

EX-OFFICIO MEMBERS

- Dr. Frank N. Beckles, Director, National Center for Family Planning Services, Health Services and Mental Health Administration, Room 12A56, Parklawn Building, 5600 Fishers Lane, Rockville, Maryland 20852.
- Col. Marcel E. Conrad, MC, USA, Director, Division of Medicine, Walter Reed Army Institute of Research, Washington, D.C. 20012.
- Dr. Howard W. Kenney, Regional Medical Director, Dept. of Medicine & Surgery, Veterans Administration, Washington, D.C. 20420.
- Dr. Robert L. Ringler, Deputy Director, National Heart & Lung Institute, National Institutes of Health, Bethesda, Maryland 20014.

[Press release, Tuesday, Nov. 9, 1971]

PROGRAM FOR EXPANDING FEDERAL ACTIVITIES DIRECTED AGAINST SICKLE CELL DISEASES

National Heart and Lung Institute, National Institutes of Health, U.S. Department of Health, Education, and Welfare, Bethesda, Maryland

A program for expanding federal activities directed against sickle cell disease, in which both community-service aspects and research will receive emphasis, has been recommended by the HEW Sickle Cell Disease Advisory Committee in their second report to HEW Secretary Elliot L. Richardson.

Sickle cell disease, a painful and life-shortening inherited disease found almost exclusively among black people, is a major health problem here and abroad. The Committee, named by HEW Secretary Richardson to advise on objectives and priorities for the program which will involve NIH, HSMHA and other agencies of the federal government, met for the second time at the National Institutes of Health October 7 and 8.

The recommendations of the Committee, which is chaired by Mrs. Ruth L. Aikens, Associate Director for Health, National Urban League, New York City, are concerned with goals and functional components of the program and allocation of the additional \$5 million recently targeted by President Nixon for sickle cell disease.

The committee recommended—

That \$2.5 million be allocated for the establishment of up to five comprehensive research and community service centers. Each center would be organized around ongoing programs in sickle-cell disease, and would bridge the gap between fundamental research, clinical application, and community service.

That \$1 million be allocated for the establishment of from 10 to 20 model screening and education clinics in various regions of the country for the purposes of (1) screening, (2) definitive diagnosis of SCD, (3) education of the population at risk and of health personnel, (4) referral of patients with SCD to appropriate sources of therapy and followup care, and (5) acquisition of detailed data on methodology.

That \$1.5 million be allocated for basic and applied biomedical research into the nature and treatment of sickle-cell disease.

The committee further recommended—

That educational and informational materials be prepared, tested, and made available through a centralized clearing house for sickle-cell disease information.

That training of black physicians, scientists, and allied health personnel in all aspects of sickle cell disease be encouraged.

That the Department of Defense screen all black recruits for sickle-cell disease.

That a survey of Federal programs be conducted for the purpose of identifying those that offer potential for assisting the sickle-cell disease program in achieving its goals.

In his February health message to Congress, President Nixon identified sickle-cell anemia as a high-priority disease target and called for a \$5 million increase in federal expenditures on sickle-cell disease during the current fiscal year. The National Heart and Lung Institute was assigned responsibility for coordinating the joint efforts of government agencies; and Dr. Robert L. Ringler, Deputy Director, NHLI, was designated coordinator of the SCD Program. The NHLI appropriation contains the FY 1972 funds earmarked for this special program, but some of these funds may be made available to other agencies to carry out part of the program.

The Sickle-Cell Disease Advisory Committee appointed by the Secretary, DHEW is composed of eleven distinguished professional and lay leaders especially knowledgeable about the problems of sickle-cell disease or active in black community programs.

BACKGROUND INFORMATION—SICKLE CELL DISEASE

Sickle cell disease, the most common inherited disorder in the U.S., is believed to be present in more than 2 million black U.S. citizens. Although the disease occurs in a relatively benign "carrier" form in most of these 2 million, approximately one in every 500 black infants is born with the life-threatening form of the disease called sickle-cell anemia. Such individuals undergo painful episodes called sickle-cell crises.

The disease is due to a genetically determined defect in hemoglobin, the respiratory pigment in red blood cells which enables them to carry oxygen throughout the body. Under conditions of diminished oxygen supply or physical or emotional stress, the abnormal hemoglobin molecules become attached to one another in such a way as to distort the normally doughnut-shaped red cells into a distinctive crescent or sickle shape.

The inability of these sickled cells to pass freely through smaller blood vessels creates a "log jam" effect, blocking the flow of blood to surrounding tissues, thus causing the extremely painful sickle-cell crisis.

Because sickle-cell disease is genetically determined, there is no way known at the present time to cure the disease in affected individuals. Current methods of treatment are aimed primarily at alleviating the pain. However, new forms of treatment may make possible reversal of the sickling process, thereby shortening or even preventing the sickle cell crises and the resultant tissue damage. These must undergo further development and evaluation before their potential can be realized.

Detailed descriptions of program components recommended by the Sickle Cell Disease Advisory Committee and a list of committee members are attached.

Additional information for potential applicants may be obtained from: Miss Rosalie J. Silverberg, Sickle Cell Disease Branch, Building 31, Room 5A-31, National Heart and Lung Institute, National Institutes of Health, Bethesda, Md. Phone: (301) 496-1187.

SICKLE CELL DISEASE PROGRAM COMPONENTS

1. Comprehensive Centers of Research and Service in Sickle Cell Disease

A limited number of Comprehensive Centers, probably no more than five, will be established on a competitive basis and supported by a special grant mechanism. Centers are conceived as identifiable units within the sponsoring institutions. Preferably, each one should be built around an ongoing program of research or service in sickle cell disease, and should be in a position to work cooperatively with community organizations concerned with sickle cell disease. Since the purpose in establishing Centers is to focus resources, facilities, and manpower in a coordinated approach to sickle cell disease problems, the program of each Center must include all of the following components: research, clinical applications, screening and counseling, education, and training of professional and allied health professions personnel. Hospital beds for clinical investigations must be available. Centers should be able to bridge the gap between fundamental research and clinical application, and promptly translate the results into demonstrations of community service.

2. Screening and Education Clinics

The Program will provide support by contract, awarded on a competitive basis, for a limited number of Clinics, probably no more than 10-20. Such Clinics will be established as demonstrations of the type of service organization capable of providing the following: education of the public as well as physicians and allied health professions personnel, screening (of 10,000 to 20,000 persons per clinic), definitive diagnosis by electrophoresis, and counseling for those found to be positive. Each Clinic should be in a setting with a referral capacity for appropriate followup and adequate medical care. Emphasis in these Clinics will be on developing methodologies best suited to mass efforts.

3. Collaborative Research and Development

In areas where specific clinical investigations or specific targets can be clearly defined (e.g. clinical trials of therapy, development of new and improved therapy, certain topics in fundamental and clinical research), work will be supported through research contracts awarded on a competitive basis. When the goals for specific individual programs have been determined, with the advice of consultants, the requirements for participation will be outlined in a Request for Proposal which will be widely circulated. An investigator feeling he can meet the specific requirements may then submit an application.

4. Biomedical Research

Research projects planned and proposed by individual investigators in accordance with their own personal interests will be supported through the traditional research project grant mechanism of the National Institutes of Health, with review by appropriate Study Section and National Advisory Council. Ap-

plications may be submitted at any time, but review occurs on a regular schedule three times a year.

5. *Training of Professional and Allied Health Professions Personnel*

Training is an essential part of the sickle-cell disease program. However, it can be carried out best in the setting of Comprehensive Centers and Screening and Education Clinics, or as part of broader programs of training in hematology or genetics. It is not anticipated that training programs devoted solely to sickle cell disease will be established, but rather that training will be an integral part of the Centers and Clinics programs described above.

6. *Public and Professional Information*

Educational and informational materials will be prepared and tested, and then made available through a centralized clearing house yet to be established. This will be a focal point for all types of sickle cell disease information.

Dr. ZAPP. Mr. Chairman, we will be glad to answer any questions the committee may have.

Mr. ROGERS. Thank you very much.

Mr. Symington?

Mr. SYMINGTON. Thank you, Mr. Chairman.

Dr. Zapp, I want to get straight, there is a recommendation for the creation of an institute within the NIH to study this problem?

Dr. ZAPP. No, we do not support the creation of an institute. We feel that the arrangement we have with the National Heart and Lung being the lead agency, coordinating research in other institutes, is the approach we would prefer to that.

Mr. SYMINGTON. I notice, for example, that Dr. Murayama worked under another institute, Arthritis and Metabolic Diseases. Is there a greater relevance of effort in that Institute to sickle cell anemia than heart and lung?

Dr. ZAPP. No. And I will turn this over to Dr. Ringler. I think there was a great personal interest on Dr. Murayama's part at a time when I think all of us in the health community, in the Government and elsewhere neglected this disease. Probably a great deal of his words was generated at his own initiative at that time within the NIAMD. I will ask Dr. Ringler if he has anything to add to that.

Dr. RINGLER. Dr. Murayama is, indeed, in the Arthritis Institute. Problems relating to the anemias at NIH are divided between the Arthritis Institute and Heart and Lung Institute.

Speaking generally, the Arthritis Institute is concerned more with understanding the basic structure and metabolism of the hemoglobin molecule.

As you know, Dr. Murayama's work has involved working out the definition of the precise structure of certain hemoglobins. Our Institute, the Heart and Lung Institute, supports the national blood resources program, which is concerned more with the clinical end of the spectrum. We are also concerned with problems of blood banking, distribution and storage of blood, and related matters.

Mr. SYMINGTON. Have studies of this kind been made of blacks in other parts of the world?

Dr. RINGLER. In terms of frequency of the disease?

Mr. SYMINGTON. In terms of frequency or treatment, or any aspect of it?

Dr. RINGLER. Information is available for other population groups. In India, for example, the frequency is much greater than it is in the United States. This is true also in areas of Africa.

As the chairman mentioned this morning, around 7 or 8 percent of the blacks in this country are carriers of the trait. In India there are groups where it is as high as 45 percent.

Mr. SYMINGTON. Because the death rate is as high as it is before 20 and 100 percent at 40, I would think the Indians would be paying a great deal of attention to it. Wouldn't their experience indicate that nutrition has something to do with it, or not?

Is this genetically related to nutrition somewhere down the line in the past or is it something that can be passed along regardless?

Dr. RINGLER. Nutritional problems would aggravate sickle cell anemia, because they contribute to the ravages of any anemia. Nutritional problems, problems of infection, all of those are greatly enlarged with sickle cell anemia.

Dr. BECKLES. Congressman, I think the historical phenomenon you are trying to explore has been demonstrated. This abnormality and the genetic pattern is very closely thought to be a normal, natural mutation of the gene.

In those areas of the world where malaria ravaged populations, this genetic mutation permitted carriers of the gene to survive where malaria was normally ravaging the population. So this is a proven phenomenon that protected millions of people many years ago.

Now that malaria is being conquered and especially in the United States where malaria has been eradicated, this genetic inheritance is no longer a necessary protective mechanism and, therefore, now needs to be eradicated or coped with itself.

Mr. SYMINGTON. Is it possible in a simple way to describe how the sickle cell blocked the effects of malaria?

Dr. BECKLES. The malaria organism lives and develops in the red cell. People who carry the trait of this disease have found the chemistry of the red cells do not support the development of the malaria organism as people with normal red cells do. This is overly simplified.

Mr. SYMINGTON. I thank you for that, Doctor. This would help explain why this disease is rampant in areas that haven't had other more sanguine approaches to the conquest of disease. That is all I have for now.

Thank you, Mr. Chairman.

Mr. ROGERS. Could I ask you did those who had sickle cell anemia but did fight off malaria still die of sickle cell anemia, or did it kind of balance off, or do we know?

Dr. BECKLES. When a person has a full-blown anemia, these people die, not necessarily from the anemia, but because of the total lowered resistance, the susceptibility to numerous other forms of disease. But the person with the trait who does not normally show symptoms is also protected and, hence, is not at the same disadvantage as the person with the anemia.

Mr. ROGERS. Has any research been done on seeing if malaria would have some curative features to it?

Dr. RINGLER. The means for controlling malaria, as I am sure you know, are quite well advanced. So, one would not want to use sickle cell anemia as a defense mechanism against malaria.

Mr. ROGERS. I ask the opposite.

Dr. RINGLER. In the opposite case, I believe the explanation goes something like this—cells that contains a high fraction of the abnormal

hemoglobin, namely, hemoglobin S, have a much shortened period of survival within the body. Normal red blood cells survive in the neighborhood of 120 days. Red blood cells in individuals with sickle cell anemia survive approximately 10 or 12 days.

Going back to Congressman Symington's question, the malaria organism, as Dr. Beckles has said, attacks the red blood cell. When the red blood cell contains a predominance of hemoglobin S, the cell is destroyed much more quickly. This provides the mechanism for getting rid of the malaria organism.

The great increases in sickle cell trait in this population come about because those individuals with the trait survived malaria whereas the others did not.

Mr. ROGERS. Thank you.

Mr. Carter?

Mr. CARTER. It is sort of survival of the fittest because of the sickle cell trait?

Dr. RINGLER. Yes.

Mr. CARTER. Very interesting. I have not heard of this theory.

I have noticed the sickle cell under a cover glass when they were deprived of oxygen.

Dr. RINGLER. That is the basis of one of the screening methodologies.

Mr. CARTER. It should not be difficult that way to find these people; is that right?

Dr. RINGLER. That particular method of screening requires a physician or someone who is rather expert. It generally is not used on a mass basis because it is subject to misinterpretation.

Mr. CARTER. You will probably miss some. You will pick them up more likely during a crisis.

What method do you use for detection of the sickle cell trait?

Dr. RINGLER. There are three categories. The one that is used most widely is a commercial test which requires that one drop of blood be placed in a test tube. If the test is positive, by that I mean if the blood contains hemoglobin S, the solution becomes cloudy or turbid. You normally hold it up against a piece of paper that has lines on it. If you can't see the lines, by definition it is positive. This is the test that is used most widely in this country.

Other tests are based on the phenomenon that you mentioned, Dr. Carter. There are tests that are suitable for automation, and, hence, can be used to screen large numbers of people. The difficulty with these is that the equipment tends to be more expensive, and an intermediate level of technical training is required in order to perform the test.

The third category of test is referred to as the electrophoretic method which detects the mobility of the various hemoglobins under the influence of an electric current. These methods require a fairly high degree of expertise in terms of technical supervision and hence are, generally speaking, not widely used for initial screening procedures.

There is one exception to that. One electrophoresis procedure that is being used in some centers, I believe is being tested here at Howard by Dr. Scott's group.

Mr. CARTER. I would like to ask you if there is any progress which has been made in the treatment of the people with sickle-cell anemia?

Dr. RINGLER. Considerable attention has been given to attempts to

shorten and possibly even reverse the sickling phenomenon. The drug which has been most widely used during the past years or more is the drug urea. It is being tested at the moment.

Our institute is supporting a clinical intervention trial to test the effectiveness of urea for this purpose. Dr. Zapp mentioned that we had awarded six contracts last spring.

Other agents are possible through a reaction that is referred to as carbamylation, which would make it possible to block the molecule and thereby would prohibit sickling. These have not reached the point of clinical application at this present time.

Mr. CARTER. What promotes the sickling? Is it the lack of oxygen?

Dr. RINGLER. Fundamentally this would be a major factor.

Mr. CARTER. Have you tried the hyperbaric treatment in cases like that?

Dr. RINGLER. I think approaches along those lines would include attempts to prevent circumstances where there would be diminished oxygen, circumstances that come about as a result of infection, pulmonary problems or situations of this kind.

Mr. CARTER. Really, then, treatment with concentrated glucose solutions and with urea and other agents have not been successful; is that correct?

Dr. RINGLER. I would not say that, sir. I would say they do need evaluation.

Mr. CARTER. In the future we possibly can develop a treatment that will be successful?

Dr. RINGLER. Yes. I would be optimistic on two bases, one would be the possibility but it has not been evaluated clinically, of using oral urea as a prophylactic treatment to prevent the crisis in the first place.

The other approach would be to find a drug that would chemically block the hemoglobin molecule so that it could not sickle. Chemically speaking, that is possible. The ability to do it in a patient has not been perfected at this time. I would reflect some optimism.

Mr. CARTER. Could you explain the mechanism of the action of urea?

Dr. RINGLER. It is thought that hemoglobin S molecules agglutinate in such a way that they assume a long, crystal structure, and that urea changes the ionic nature of the cell, or the electrical property of hemoglobin within the cell, in such a way that these molecules do not agglutinate.

Mr. CARTER. They are repelled rather than attracted?

Dr. RINGLER. Yes. There is a change in the hydrophobic nature of the contents of the cell.

Mr. CARTER. Basically, though, at the present time you think that our efforts should be pointed toward screening these people to find the genes that carry the sickle cell trait and advise and counsel them about marrying?

Dr. RINGLER. The attack should perhaps be two-pronged. One major segment of it should be directed at counseling and prevention of the disease in the sense that you just used it. Perhaps the second prong should be directed at perfecting better therapy for those who already have the disease. I think these are the two major approaches to the problem.

Mr. CARTER. What are the final stages of this disease? Could you explain that?

Dr. RINGLER. I think the final cause of death——

Mr. CARTER. What happens prior to death?

Dr. RINGLER. I think that perhaps most frequently the cause of death is associated with pneumonia. Other causes of death are related to infection and associated again with the ravages that come about as a result of very profound anemia.

Mr. CARTER. Do you get any embolic and thrombotic phenomena?

Dr. BECKLES. Mr. Chairman, this disease produces great mimicry. It mimics so many other kinds of diseases that it appears the cause of death might be due to a heart which has failed. It might be due to pneumonia. The mere lowered resistance to infection, the creation of thromboli or emboli and then infarction, brain damage, bone damage, liver damage in any imaginable series. There are a series of contributing causes to death in sickle cell patients.

Mr. CARTER. That is exactly what I wanted to know. Thank you.

Thank you, Mr. Chairman.

Mr. ROGERS. Mr. Kyros?

Mr. KYROS. Thank you, Mr. Chairman.

If the partners both carry the sickle cell anemia traits, but are prevented from having children or counseled against having children, would that do away with the disease entirely?

Dr. BECKLES. Hopefully. This is what we must do as a preventive measure through education and through counseling, equipping individuals to make informed judgments. We feel that with the availability of this kind of counseling, people will make responsible decisions as it affects their yet unborn progeny. But this is only one major area.

Mr. KYROS. We recently in this committee worked on a family planning services bill which is administered through your department. Does that include blood tests for blacks, or screening sickle cell anemia?

Dr. BECKLES. The Family Planning Service Centers will add sickle cell screening and diagnosis as a component of the medical service being provided to people who come in for family planning services.

Mr. KYROS. Throughout all the States in the United States, to doctors and marriage bureaus who require blood tests, so that among one of the tests they are supposed to give people who happen to be black is one of these blood tests? Do they know that? Is that information given to everybody?

Dr. BECKLES. I am not sure. I would hazard a guess that it is not being done on a nationwide basis.

Mr. KYROS. How many States require blood tests before marriage? Do we know?

Dr. BECKLES. We can supply that for the record, Mr. Kyros. I think a decreasing number of States require the blood test by law now.

(The following information was received for the record:)

STATES NOT REQUIRING BLOOD TESTS BEFORE MARRIAGE

Only five States do not require blood tests before marriage. They are: Maryland, Minnesota, Nevada, South Carolina, and Washington.

Mr. KYROS. How do we screen out Rh blood factors in women? Is it through blood tests prior to marriage, or is it voluntary by those people who are wealthy enough to check themselves before they have babies or before they are pregnant?

Dr. BECKLES. A combination of those ways.

Mr. KYROS. In this case, something must be done before conception, otherwise if the two partners are carriers of the trait, then with no question, the child does get sickle cell anemia?

Dr. BECKLES. Statistical studies of a large number of matings of persons with the sickle cell trait show the proportionate number of children to be one-fourth with normal blood, one-half with sickle cell trait, and one-fourth with sickle cell anemia.

Mr. KYROS. If a couple has four children, and both parents have sickle-cell anemia, does that mean that the four children will be affected?

Dr. BECKLES. That holds out in the ratio for each child. What one sees on occasion is that each child born will have the disease, or that each child born could be normal. In some instances two and three children in the same family will have sickle cell anemia with the disease components.

Mr. KYROS. Community screening is the way I think we should approach sickle cell anemia. Getting back to research, I would like to know why it is that for 20 years we have known about sickle cell anemia, but we haven't determined its cause.

Dr. ZAPP. I think it has been a matter of inattention, no advice or leadership from either the general health community or the Federal Government itself. I think this is the first year that there has been a combination of those factors that have resulted in bills before your committee and activity in the Department.

Mr. KYROS. In the huge cancer research program that we are legislating, will there be some provisions that will help sickle cell anemia? Will the research on blood cells in the cancer program help?

Dr. ZAPP. I would say it would be pretty tangentially related. On that specific question, I would want to refer to Dr. Ringler.

Dr. RINGLER. I am not aware of any benefit that is slated to come from that program.

Dr. ZAPP. I would guess probably it would not be directed that way. Benefits would more likely come about through the three institutes we mentioned earlier, which have sickle cell disease and related research activities going on.

Mr. KYROS. Thank you very much.

Mr. ROGERS. Mr. Symington, I believe you have one more question?

Mr. SYMINGTON. Thank you, Mr. Chairman. I want to ask if this condition can be induced in rats or mice and tested in this way?

Dr. RINGLER. It occurs in certain animals. The one perhaps most widely known is the deer. A suitable animal model, however, is not available at this time.

Mr. SYMINGTON. This is not an available area of research?

Dr. RINGLER. There is not a suitable animal model for such a study.

Dr. BECKLES. One of the members asked the question before why is there not more work in this area being done in areas of India and Africa where it has been so prevalent? I think the answer to that is that these countries have been faced with so many other problems that they have no resources to turn their attention to this particular disease. Whether or not animals there would turn out to be suitable since they are in the same endemic region is a question which has not been explored. It is an interesting one.

Mr. SYMINGTON. Thank you.

Mr. ROGERS. May I ask a few questions? Why can't your committee encourage States to require in their licensing for marriage a blood test as they do for other diseases?

Dr. ZAPP. I think on that point, Mr. Chairman, the committee has only had preliminary meetings. Their first charge was to take the money provided in this year's budget and determine the best way to use it and where our program should be going.

Mr. ROGERS. Isn't that the quickest way to get something done, right at the marriage level, immediately?

Dr. ZAPP. Yes; obviously, to prevent the occurrence of the disease is always the best route.

Mr. ROGERS. You don't know whom to counsel either.

Dr. ZAPP. I think this could very well be one of the areas where the committee would be getting involved.

Mr. ROGERS. I don't know why the thing can't go on immediately. What do we have to wait for? We know you want to counsel people if they have the sickle cell trait.

Dr. ZAPP. You certainly do; but in a full-blown program, it must be determined exactly where is the best place to intervene.

Mr. ROGERS. You are not intervening anywhere right now. So why not go ahead and encourage them? Send out a wire or a letter to every State, every public health State official, saying we would urgently request that you immediately test for sickle cell and advise those people if this condition exists when there are two about to marry. Should they at least be advised immediately?

Dr. ZAPP. Yes; they most certainly should. I see no objection to doing it.

Mr. ROGERS. This requires nothing except a letter.

Dr. ZAPP. I think there are some obvious problems related to the counseling capabilities within his area of responsibility. He gets two people who are ready to apply for a license and suddenly they are both informed they have the trait. When he tells them that he will also have to have somebody there who has some experience and expertise in being able to relate to them the problems, the possibilities, and the effects.

Mr. ROGERS. Can't you put out a little one-page sheet saying this is the problem and you should consult a physician for further knowledge?

Dr. ZAPP. I can see nothing in this that we should object to at all. I would like to see us put together a package to inform health officials about more than just one thing.

Mr. CARTER. Along this same line in the United States, at the present time how many people have diabetes? Could you give me an answer to that?

Dr. ZAPP. How many?

Mr. CARTER. Yes, sir; what percentage of the people?

Dr. ZAPP. I don't know. I think there was something in an earlier statement.

Mr. CARTER. Isn't it true that if two diabetics marry that their offspring will also be diabetic? And further diabetics really could more than likely be eliminated if one chose his partner without the diabetic trait, is that correct?

Dr. ZAPP. That is right.

Mr. CARTER. We ought to do that, too, should we not, immediately inform the public?

Mr. ROGERS. If the gentleman will permit, I don't know why we have not done it. It is unbelievable. This could be done, advise people with a simple letter while they are making the blood test anyhow.

Dr. BECKLES. I think we have gone here into an area which is not as productive, predictably productive, as one might think it ought to be. The fact is that education and information should be made available. Whether or not this would necessarily result in modification of behavior at that stage of the relationship—

Mr. ROGERS. I understand. I doubt if it will change very many. I don't say that. At least it will give them the knowledge and let them make an informed judgment and let them get some counseling.

Mr. SYMINGTON. I wonder why we have to wait until the threshold of wedlock before each partner knows.

Dr. BECKLES. This is the point I was going to arrive at. Perhaps the point of greater yield would be the earlier ages when the individuals can inculcate that knowledge and deal with it as a progressive thing. But there is a point at which one has to act, since there are people who will continue to get married who most likely carry the trait—

Mr. ROGERS. Of course the ideal situation is to find it out as we can but we simply are not doing it and \$5 million is not going to do it.

Mr. CARTER. Along the same line we might also check Rh factors involved in young ladies who are going to be wed.

We could advise them.

Mr. ROGERS. I would think you would set forth what blood test, with scientific knowledge now, could be helpful to people in preventing disease, and giving counseling. Most States require blood tests, don't they?

Now could they not run those very tests with probably almost the same amount of blood that they take now, or a little more?

Dr. BECKLES. I think once one builds a national mechanism to deal with the numbers of people so involved then the things you are inquiring about will become possible. At this point in time there is not a competent or existing national mechanism that could immediately accomplish this.

Mr. ROGERS. That is why you are going to have to require the States to do it, or encourage them to do it.

Dr. BECKLES. Yes.

Mr. ROGERS. But I haven't seen any letters from HEW saying we would urge or I haven't seen any funding to encourage such a program in the States. Now this might be helpful.

Is it good to know that you have diabetes? She may not know it. Is it good to know that? Can you do something about it?

Mr. CARTER. It is not whether she has it but whether her parents had it.

Mr. ROGERS. I mean when they are getting ready to marry. I thought you said if both had it, then the child would get it. Aren't there things that can be done if you find out you have it?

Dr. BECKLES. Mr. Chairman, you are entering another area, namely the philosophy of prevention rather than treatment.

Mr. ROGERS. Which is about time.

Dr. BECKLES. I agree but in this Nation prevention as a concept toward medicine has not been a national—

Mr. ROGERS. This is what we are trying to change. This committee is committed to this.

Dr. BECKLES. I see the thrust of your question.

Mr. ROGERS. It is trying to get some treatment that will prevent as well as rely on just curative medicine.

Mr. KYROS. Mr. Chairman, will you yield for a question?

Mr. ROGERS. Certainly.

Mr. KYROS. When babies are born, black or white, is there any reason to give them any blood tests?

Do they normally give newly born children a blood test for anything now?

Dr. RINGLER. Under some circumstances what is called cord blood is taken.

Mr. KYROS. Would it not be one way to start, from the very beginning when the child is born?

Dr. BECKLES. Methodologies are being developed now to find out at what point in time one can most easily identify the disease. The sickle cell hemoglobin is masked at birth, and therefore this is not the most appropriate time. Research is now being done to discover how soon after birth one can predictably make a true test.

Mr. KYROS. We don't know from zero to puberty?

Dr. BECKLES. One can do this somewhat around 6 months now but hopefully it could be done eventually.

Mr. KYROS. Thank you.

Thank you, Mr. Chairman.

Mr. ROGERS. I would hope that you would seriously go into this situation of at least encouraging the State people to do this testing with the blood samples they require anyhow. I imagine most of them would respond.

Dr. ZAPP. I find it inconceivable, Mr. Chairman, with the advisory committee we have, which is acting pretty rapidly, having held its first meetings on the 13th of August and with very dedicated, interested and knowledgeable people on this, to think that this is not one of the kinds of things they are going to look at. We are looking at a problem that people have had for years. As far as responsible individuals in Government—

Mr. ROGERS. I am not condemning the fact, it is fine you are moving, I am not deprecating that. What I am saying is we know this is one technique that could be quickly available. I don't see that any committee has to rule on it or discuss it for 3 or 4 months. You have enough judgment within NIH to say this ought to be done. I think Dr. Carter's suggestion of these other blood tests might be helpful, at least make the suggestion that this be incorporated in the blood test given at marriage. This is at least one route to get a lot of people tested on these various diseases.

You may get some feedback, to do this we need another \$100,000. Well, we ought to know that. This committee ought to know it. Maybe we can be helpful. So I would urge you to do that.

Mr. CARTER. Mr. Chairman, it is commonly known that a diabetic marriage, both of them having diabetes, their off-spring will have it. That is common knowledge. Perhaps it has not received the emphasis that it should have received.

Dr. ZAPP. I certainly think in previous years it was an almost untouchable subject to discuss who should and who should not marry and for what reasons or to discuss the genetics of the races in our country. It was not something that was public domain. Certainly, I think the discussions that have occurred today have brought out some points. To my knowledge, there has been no program of diabetic counseling, even though you say we all knew about it. We ought to take a look at what has the highest priority and what is the best setup to do these things.

Mr. CARTER. Again, the Rh factor would certainly be helpful.

Mr. ROGERS. I think these suggestions are excellent. Now, why don't we incorporate that? Let us know right away, will you, if your department will do that? If not, this committee will do it. I think you ought to do it.

Mr. KYROS. Mr. Chairman?

Mr. ROGERS. I will yield.

Mr. KYROS. Dr. Zapp. I have been listening to you this morning, explaining what we have failed to do for 20 years through oversight and inattention. We are now allocating \$5 million, but the Sickle-Cell Disease Advisory Committee advises the Secretary of HEW that \$5 million is not enough and that we should go to \$25 million. Do you agree?

Dr. ZAPP. No; the order was reversed. The initiative on the President's part came prior to the appointment of the committee. There was a limited \$1 million-plus sickle-cell disease research program in the Department. Some other peripherally related programs or activities were going on around that time. The committee was appointed in August of this year after the appropriation procedure had been completed. They were not in the position to deal with the issue of the amount in the budget.

Mr. KYROS. I think that what the President has done to highlight this disease is enormously commendable. It shows he is paying great attention to it at this time. Does the Sickle-Cell Disease Advisory Committee feel that we can't absorb a rate of spending of \$25 million? We have discussed in the cancer bill that sometimes money cannot be spent anyway even when it is poured out. What would happen if you did have the \$25 million instead of \$5 million? Would we move forward more rapidly in the research and screening of this disease?

Dr. ZAPP. On the research side, I think the question would probably focus first on how much the research capacity can be accelerated at one time; is it two, three, four, or five times in a year over what you have been previously doing? I think any progress there would be limited for a variety of reasons, but mainly by the need to have competent researchers who are involved and on the spot. In other areas, there is the choice of what is the best mechanism. Do you put everything together and say that you will fund screening in all available health centers and all maternity centers and youth centers and everywhere else in the country, or do you use a certain amount of that money for education and counseling, setting up the kind of methodologies that are necessary to make sure they are part of all the ongoing programs? Frankly, at this point, that is what we are grappling with.

Mr. KYROS. Dr. Connally at Johns Hopkins Hospital has said that

he believes \$25 million a year would be well spent on sickle-cell anemia. He has stated that for the record.

Dr. BECKLES. Mr. Congressman, the committee has not dealt with increased sums of money. I don't think it is fair to project what the committee would say if asked to deal with increased sums. They have dealt only with the \$5 million, even though they have expressed feelings that there could be more money spent in this area.

Mr. KYROS. I am impressed by the fact that the test can be administered at 0.02 cent to 0.08 cent per sample. It takes millions of dollars to care for those who have the disease, yet, the tests to discover it cost only 0.02 cent to 0.08 cent a sample.

Dr. ZAPP. This depends on the method used. There is a method that, it is thought, would give results at that price. This is the one that would call for high levels of skilled technicians to perform.

Mr. KYROS. Thank you, Mr. Chairman.

Mr. ROGERS. I am anxious to pursue this business of simply a letter going out from HEW to all your State departments of health urging and requesting that they incorporate these tests into the blood testing for a marriage license. Would you let us know if you will do it; and if not, this committee will see if we can.

Dr. ZAPP. Yes, sir.

DEPARTMENT OF HEALTH, EDUCATION, AND WELFARE,
OFFICE OF THE SECRETARY,
Washington, D.C., February 24, 1972.

HON. PAUL G. ROGERS,

*Chairman, Subcommittee on Public Health and Environment, Committee on
Interstate and Foreign Commerce, House of Representatives, Washington,
D.C.*

DEAR MR. ROGERS: During the November 12, 1971 hearings before your Subcommittee on H.R. 11171, "a bill to provide for the prevention of sickle cell anemia," you asked whether or not the Department of Health, Education, and Welfare would consider urging the states to incorporate into the blood tests required of applicants for a marriage license, a test for sickle cell anemia and possibly other genetically transmitted diseases. We are, as indicated in our testimony before the Subcommittee, interested in sickle cell disease as well as other genetically transmitted diseases. As you know, the Department in Fiscal Year 1972 committed \$10 million to the sickle cell initiative and intends to expand the program in Fiscal Year 1973 to \$15 million. I believe these actions indicate that our commitment to do something about this neglected disease is sincere.

On the question of urging states to undertake more expanded sickle cell testing activity, there are a number of issues to be considered, each one of which could have a significant impact on the success of any such program. One such consideration is the usefulness of performing this test at this particular time in an individual's life. We hope it will be possible for persons to know earlier in their lives if they have the sickle cell trait and research is currently being done to find a more appropriate time for such testing. Other important considerations are the availability of personnel and equipment to perform and evaluate such testing, as well as follow-up services for those found to be carriers of the sickle cell trait. There are also possible legal constraints; e.g., confidentiality of test information, as well as the need to evaluate other similar such activities which may have been taken in the past.

Very frankly, we do not now know what the detailed procedures are in the forty-five states which require a blood test before they will issue a marriage license. We are, therefore, forwarding the text of the enclosed letter to the State Governors asking for their assistance in this area. We believe this letter will stimulate significant activity at this time and provide us with information on

the issues described above. It is our expectation that we can resolve these questions and be able to report a course of action to you within four months.

Your interest in sickle cell and other genetically transmitted diseases is appreciated and we will keep you informed of our progress on this effort.

With kindest regards,
Sincerely,

ELLIOT L. RICHARDSON, *Secretary.*

DEPARTMENT OF HEALTH, EDUCATION, AND WELFARE,
OFFICE OF THE SECRETARY,
Washington, D.C., February 24, 1972.

HON. NELSON A. ROCKEFELLER,
Governor of New York,
Albany, N.Y.

DEAR GOVERNOR ROCKEFELLER: Sickle cell trait and sickle cell anemia are inherited disorders caused by a genetically determined abnormality in the hemoglobin of red blood cells. The best available figures suggest that about 2 million black citizens carry sickle cell trait. They have no symptoms except under the most unusual circumstances and may not even be aware that they carry the trait. However, carriers of the trait can pass the disorder on to their children and if they happen to choose a mate who is also a carrier of the trait, there is a one in four chance that each child will inherit the trait from both parents and thus will have sickle cell anemia.

It is estimated that one in 500 black infants is born with sickle cell anemia and that a total of 50,000 black citizens have sickle cell anemia, a painful, debilitating and life shortening form of the disease. The condition is found primarily among black individuals, but it has been noted occasionally in Caucasians of Mediterranean extraction, Spanish-speaking populations, and populations from the Far Eastern subcontinent. The fundamental chemical basis for the disease and the manner of inheritance are well understood. Because of its genetic nature, there is no known cure at the present time and it is unlikely that a cure can be found in the foreseeable future. However, expanded programs in screening and education can provide affected individuals, particularly carriers of the trait, with the knowledge necessary to make informed decisions regarding their own lives.

In his Health Message in February 1971, President Nixon specifically noted the need for expanded services and research programs in sickle cell disease. Five million dollars was appropriated to HEW to implement this initiative in Fiscal Year 1972. This amount was added to approximately \$1 million already being spent on sickle cell disease research. The Department subsequently increased the total for this year to \$10 million, and the President is recommending that \$15 million be made available for these activities in Fiscal Year 1973.

A Sickle Cell Disease Advisory Committee composed of eleven distinguished professional and lay leaders especially knowledgeable about the problems of sickle cell disease or active in black community programs was appointed to advise the Department on objectives for the program and on priorities and plans for implementation. Based upon their recommendations, the HEW program now being implemented includes:

1. The establishment of approximately 5 comprehensive centers of research and service.
2. The development of approximately twenty model screening and education clinics.
3. The implementation of expanded programs of basic and applied research.
4. The establishment of a clearinghouse for all types of sickle cell disease information.

While these efforts cannot alleviate all the problems associated with sickle cell disease, we do feel that they are a productive first step in this area. Accordingly, the Department wishes to coordinate these endeavors in the most effective manner with other ongoing activities.

Sickle cell represents just one of several diseases and disabilities which are transmitted genetically from parent to offspring. It is assumed from information available to this Department that many of these diseases are not included on a regular basis in screening procedures required or recommended by most states.

It is, therefore, the purpose of this letter to ask you to provide to the Sickle

Cell Disease Program Coordinator, Sickle Cell Disease Branch, National Heart and Lung Institute, National Institutes of Health, Bethesda, Maryland, 20014, information concerning what is now being done in the way of sickle cell screening programs in your state, including any requirements for testing at the time of issuance of marriage licenses, what methods are being used, and what further plans you are developing for sickle cell screening programs. It would be appreciated if we could hear from you within sixty days.

With kindest regards,

Sincerely,

ELLIOT L. RICHARDSON, *Secretary.*

Mr. ROGERS. You had a meeting of your committee, and one of the things I understand from your testimony here is that they were going to think of something else the Federal Government could do which could be done immediately; that is, have the Defense Department give testing. Has that been done?

Dr. ZAPP. They have made the recommendations. Of course, the recommendations and press release on that came out only the other day. I think their meetings were October 7 and 8, at which time their recommendations were formulated.

Mr. ROGERS. The reason I ask that is that you end up your statement by saying you can't speak for the Defense Department and Veterans' Administration.

Dr. ZAPP. As an official of the HEW, I can't speak for the commitments of the Defense Department or Veterans' Administration. It is something they have to answer for themselves.

Mr. ROGERS. This is in the health field. That is one of the problems, trying to get someone to speak for health.

Dr. ZAPP. Well, being able to speak and being able to allocate human and physical resources are something different. The other thing I mentioned is that we did want to bring before the committee the complete recommendations of our advisory committee, and these were two of them.

Mr. ROGERS. Is there a time limit as to when they are to be executed? If you can't speak for the Department of Defense, how do we know? They may not agree with you.

Dr. ZAPP. I think the bills before this committee have similar provisions in them.

Mr. ROGERS. Then you think we have to do it by law?

Dr. ZAPP. No; I am not saying that. You may want to get that answer from them.

Mr. ROGERS. I thought you had already called them in and they were in this committee?

Dr. ZAPP. They are represented on this advisory committee, but to say they can come in overnight and say that the Secretary of Defense or the Administrator of the Veterans' Administration has consented to this program and they are ready to implement it, would be impossible. I do think the gentlemen with me who are members of that committee can give you a better flavor of what they consider to be the possibilities.

Mr. ROGERS. Even that would be helpful.

Dr. RINGLER. There is an ex-officio representative from the Department of Defense on the committee. There is also a representative from the Veterans' Administration on the Committee. At the last meeting the committee recommended that all recruits for the military

service be screened and that all the patients that are cared for by the Veterans' Administration be screened. This was discussed at the committee meeting.

These recommendations will be transmitted to both agencies. It is my informal impression that the agencies are receptive to those recommendations.

Mr. ROGERS. I would like the counsel to contact those agencies right away and let us get their reaction to see if they are going to do it. If not, we will put it in the law if that is the only way to get that.

I think your recommendations are good and I am glad you got them on the committee.

Mr. SYMINGTON. Will the chairman yield?

Mr. ROGERS. Yes.

Mr. SYMINGTON. It occurred to me that there are problems involving unwed mothers that have no intention of marrying. It seems curious that when we call something a social disease we are willing to insure a test being made. Thus the Wasserman test is well known. I wonder why we discriminate in favor of the Wasserman test when there are other killers like this.

Perhaps there ought to be a complete catalog of killers which in legal combination between marrying partners or nonmarrying partners, as the case may be, can produce a defective child. No one wants to do that. It seems to me that perhaps there is a mission for the Public Health Service to get the word out, a whole series of things of this kind, so that people know what they are experimenting with.

Dr. BECKLES. Mr. Symington, I think we have made initial steps in that direction by attempting to combine the resources of the National Institutes of Health, the Health Services, and Mental Health Administration, Social Rehabilitation Service, and Social Security Administration, to bring in the variety of agencies which touch the various strata of the Nation so that, indeed, for education and then action, we can get the total Government mechanism into this effort.

Mr. SYMINGTON. Thank you.

Mr. ROGERS. I would like to have for the record the number of research contracts that have been let, that you plan to let, and the time limit, with the \$5 million.

(The following information was received for the record:)

ALLOCATION OF FISCAL YEAR 1972 FUNDS EARMARKED FOR SICKLE CELL DISEASE

On the recommendation of the Sickle Cell Disease Advisory Committee, the \$5 million appropriated for sickle cell disease will be allocated among the following Program Components:

1. \$2.5 million for up to five Comprehensive Centers for Research and Service, which will address all aspects of the Program with about 40% allocated to research aspects and about 60% to community service aspects;
2. \$1.0 million for up to 10-20 Screening and Education Clinics designed primarily to develop appropriate methodologies before mass screening programs are undertaken;
3. \$1.0 million for Collaborative Research and Development, i.e. target-oriented research addressing the biological problems of the disease;
4. \$0.5 million for expansion of support of research through the traditional research project grant.

In addition, an Information Center and clearing house will be established. If additional funds are allocated for sickle cell disease this fiscal year, the approximately equal balance between research and service aspects of the Program, as recommended by the Advisory Committee, will be maintained.

At the time of this writing, none of the earmarked funds have been spent. However, Requests for Proposal and/or guidelines and application forms have been sent to all individuals and groups who have requested specific information on one or more of the Program components. Applications are now being received for late winter and spring review, with funding anticipated in May and June. The present status of the major components is shown in the attached table. Traditional research project grant applications are received and reviewed on a regular schedule three times a year.

PRESENT STATUS OF MAJOR COMPONENTS OF
SICKLE CELL DISEASE PROGRAM

	<u>CENTERS</u>	<u>CLINICS</u>	<u>COLLABORATIVE</u>
Administrative responsibility for component	NIH	HSMHA	NIH
Support mechanism	Special grant	Contract	Contract
Announcement or Guidelines Available for mailing	11-8-71	1-7-72	11-19-71
Indications of intent to submit proposal	approx. 20	approx. 100	approx. 100
Deadline for submission of proposals	2-1-72	2-18-72	1-14-72
Number of applications received			approx. 60

Mr. ROGERS. I got the gist from the testimony that you are not doing so much basic research as you are more simple treatment and research on how to define more readily the disease. Am I correct in that?

Dr. ZAPP. I think there is both some targeted and some basic research. Obviously the basic research takes place in a variety of institutions.

Mr. ROGERS. How much money would you say is being devoted, of the \$5 million, to basic research?

Dr. RINGLER. None of the \$5 million has been spent as of today.

Mr. ROGERS. None has been spent?

Dr. RINGLER. As of today. We waited before making awards until we received the advice of the committee. The committee recommended that about half of the \$5 million be used for research and half be used for community service.

In answer to your last question, I would estimate that approximately half a million dollars will be spent for basic research.

Mr. ROGERS. What would be the other research being done?

Dr. RINGLER. The other would be of a more clinical nature, attempting to evaluate and improve the therapy for sickle cell crises and developing better methodology for managing these patients.

Mr. ROGERS. How much basic research are we doing in cystic fibrosis?

Dr. ZAPP. We will have to supply that for the record.

Mr. ROGERS. Do you know how much for muscular dystrophy?

Dr. ZAPP. No.

(The following information was received for the record:)

Cystic fibrosis research—NIAMD

1968	-----	\$2,001,000
1969	-----	2,588,000
1970	-----	2,507,000
1971 estimate	-----	2,643,000
1972 estimate	-----	2,817,000

Muscular dystrophy research—NINDS

1968	-----	\$3,899,000
1969	-----	5,139,000
1970	-----	5,092,000
1971 estimate	-----	5,476,000
1972 estimate	-----	5,337,000

Mr. ROGERS. In other words, you are planning about a half million dollars on basic research?

Dr. RINGLER. May I make one more statement? Your question was with regard to the \$5 million?

Mr. ROGERS. Yes.

Dr. RINGLER. In addition there is the sixth million. That is all in the area of basic research.

Mr. ROGERS. All \$6 million in basic research?

Dr. RINGLER. No, \$1 million.

Mr. ROGERS. One million dollars plus the \$5 million?

Dr. RINGLER. One million dollars plus approximately a half million dollars from the \$5 million.

Mr. ROGERS. So the total of basic research of \$1.5 million?

Dr. RINGLER. Approximately.

Mr. ROGERS. How many researchers do you have working on that?

Dr. RINGLER. I can't give you the number.

Mr. ROGERS. Do we have an estimate? Ten, five?

Dr. RINGLER. I would estimate it in the neighborhood of 30 or 35 grant programs.

Mr. ROGERS. Will you let us know what those are?

Dr. ZAPP. We can submit that exactly.

Mr. ROGERS. I think that would be helpful.

(The following information was received for the record:)

RESEARCH GRANTS RELATED TO SICKLE CELL DISEASE

In fiscal year 1971, the last year for which we have complete figures, there were 22 research grants with primary emphasis on sickle cell disease and at least 25 in which a component was related to sickle cell disease.

Mr. ROGERS. Also how many people would be screened in these clinics you speak of, up to five clinics. Do we have any planning for that?

Dr. RINGLER. The number is in the range of 10 to 20 clinics.

Mr. ROGERS. I don't think so. Not according to your testimony. Maybe I misunderstood. I thought you said five.

Dr. BECKLES. There are two approaches proposed.

Mr. ROGERS. Wait, how many clinics.

Dr. BECKLES. It is proposed that 10 or 20 clinics would be funded using approximately \$1 million out of the \$5 million.

Mr. ROGERS. I see. This would be screening and education clinics, 10 to 20, out of \$1 million.

Dr. BECKLES. Yes, Mr. Chairman.

Mr. ROGERS. You feel you can set up those clinics?

Dr. BECKLES. Yes, sir.

Mr. ROGERS. With \$1 million, 20 clinics?

Dr. BECKLES. We feel that 10 to 20 clinics can be set up. These would be planning clinics or experimental clinics as opposed to a series of standard clinics across the Nation.

Mr. ROGERS. In other words, these would serve relatively few people?

Dr. BECKLES. Yes, sir.

Mr. ROGERS. It would be for demonstration?

Dr. BECKLES. Precisely.

Mr. ROGERS. Not a real functioning clinic to screen large numbers?

Dr. BECKLES. The numbers would be limited according to the funding.

Mr. ROGERS. How many would you be able to serve per clinic?

Dr. BECKLES. It depends on whether the money goes to an ongoing center where it is estimated that perhaps \$25 per patient is required for the three facets or whether it will be a brand new effort that is funded.

Mr. ROGERS. I think we need to know that.

Dr. BECKLES. We will give you an estimate of the number that would be served by this method.

(The following information was received for the record:)

NUMBER OF PEOPLE THAT CAN BE SERVED BY \$1 MILLION FUNDING
OF SCREENING AND EDUCATIONAL CLINICS

It is our estimate that 100,000 people can be served in the programmatic component of screening and educational clinics funded at a level of \$1 million.

Mr. CARTER. On that very thing, if you wanted complete screening of those people who intend to marry, it seems to me it would be most logical to do the screening through the hospitals and the State health departments throughout the country. That is the way we do our VDRL tests at the present time.

Dr. BECKLES. What we are trying to seek, Mr. Carter, is the best way to approach the problem. If one should propose to set up a series of laboratories for screening across the Nation, this would require an enormous sum of money just for facilities and equipment. We are projecting hopefully to be able to centralize the laboratory part of the effort before we put in motion a national program.

Only yesterday afternoon we talked with the Association of Neighborhood Health Centers which is comprised of seven neighborhood health centers and approximately a hundred thousand people. That question arose; namely, should you propose to put a screening laboratory facility in each neighborhood clinic. They said, no, there is one that can do the job for seven. This is the kind of approach we are trying to look for in the research phase.

Mr. CARTER. If you want to do other checks, it should be done through established laboratories?

Dr. BECKLES. This is the other component that we talked about in the nature of the centers.

Mr. ROGERS. You would have five of those centers?

Dr. BECKLES. Yes.

Mr. ROGERS. Would you give us a description of what they would do, for the record, and how many people would be affected?

Dr. ZAPP. Mr. Chairman, in supplying that for the record, we would like to offer for the record the fact that we are also preparing an inventory of on-going screening activities in the maternal youth centers and some of the other departmental centers. Because of their local orientation, some have them and some don't. This is the first time that the program has taken a look at all of these.

This material was put together on a preliminary basis for the advisory committee. The Department is in the process of getting more hard material of the type you are looking for.

Mr. ROGERS. Has the committee made all its recommendations?

Dr. BECKLES. The committee has made its recommendations to date based on the \$5 million and the work that has been presented to them.

Mr. ROGERS. Will you list for us all their recommendations?

Dr. BECKLES. Yes, sir.

Mr. ROGERS. And what action has been taken as a result of those recommendations?

Dr. BECKLES. This was included in the material that Dr. Zapp requested be placed in the record, Mr. Chairman. (See pp. 64-67.)

Mr. ROGERS. Are there any other questions? If not, thank you very much. Please let us know about this letter to the health departments.

Dr. ZAPP. Yes, sir.

Mr. ROGERS. Our next witnesses are from the Black Athletes Foundation for Research in Sickle Cell Disease, Pittsburgh, Pa. The executive director is Horace Davis; along with Henry Aaron of the Atlanta Braves; John Henry Johnson, former pro football player; and Dr. Anthony Georgio, University of Pittsburgh.

We welcome you to the committee. If you would like to take your seats at the stand, please come forward.

Mr. Davis, you are going to make a statement?

STATEMENT OF JOHN HENRY JOHNSON, FORMER PRO FOOTBALL PLAYER, IN BEHALF OF THE BLACK ATHLETES FOUNDATION FOR RESEARCH IN SICKLE CELL DISEASE; ACCOMPANIED BY HORACE DAVIS, EXECUTIVE DIRECTOR; HENRY AARON, ATLANTA BRAVES; AND DR. ANTHONY GEORGIO, MEMBER, MEDICAL ADVISORY BOARD

Mr. DAVIS. John Henry Johnson will read the statement. I will be ready to answer any questions.

Mr. ROGERS. That will be fine.

Mr. Johnson, we are delighted you are interested in the subject and the fact that you are doing something to bring public attention to a problem which has been neglected greatly.

Mr. JOHNSON. Mr. Chairman, all of us wish to offer our sincere thanks for being given this opportunity to address you on this subject of such great importance to us and to millions of other black Americans.

So that there will be time for you to ask whatever questions you may have of us, I will touch upon only a few aspects of this major health problem and the legislation concerning sickle cell anemia that is now before you.

Little more than 2 weeks ago, on October 27, our foundation presented testimony before the Subcommittee on Public Health of the Senate Committee on the District of Columbia, which was considering legislation which would establish in the city of Washington, D.C., a pilot program on sickle cell anemia. Because much of that testimony also is relevant to the bill now before this subcommittee, let me just outline the major points we then stressed:

Because statistics on the incidence of this or any other disease attain meaning and significance only to those who understand what it does in the individual case, it is important to recognize that sickle cell anemia not only involves premature death, but also untold suffering for the patient and his or her family. We described a case which followed a typical course: 18 hospitalizations by age 24 and death soon after.

A total program, involving a variety of public and private agencies, is the only kind that can lick this disease. It must be both adequately funded and properly coordinated.

Washington, D.C., the only city in the country in which a majority of residents are black, is a most appropriate site for an initial federally funded pilot program dealing with sickle cell anemia.

Testing and screening on a massive scale can, we strongly believe, drastically reduce the incidence of sickle cell anemia, although all aspects of the program must and should depend upon the voluntary cooperation of the affected communities and individuals.

Clearly there is a growing recognition in Congress of the need for action in meeting the serious problem of sickle cell anemia. In testifying today in strong support of the National Sickle Cell Anemia Prevention Act, we wish to commend those Senators and Congressmen who are sponsoring this measure and the companion bill that would establish a demonstration program here in the Nation's Capital. We urge prompt and positive action on both proposals.

It is estimated that today 50,000 Americans are dying of sickle cell anemia, a fatal disease which kills a majority of its victims before they reach 21 years of age. It is a disease that involves terrible pain and suffering. Because it is hereditary—that is, passed on in the genes of the parents to a child—it can mean tremendous feelings of guilt for the parents of its victims, who often learn of the risks involved in having children only when the disease shows up in one of their own. For the afflicted family, too, the financial burden of sickle cell anemia is staggering.

More than 2 million black Americans—one in every 10—carry the sickle cell trait. Most are symptom free, and their greatest risk lies in marrying another trait carrier and having children who have the disease. The chance is very high—25 percent in every case—that a child of two trait carriers will have sickle cell anemia. Trait carriers, too, however, may themselves experience “sickle cell crises” in certain situations where the normal supply of oxygen is reduced, such as at high altitudes or under general anesthesia. And a few, under normal conditions, may show symptoms which, while far milder than those associated with anemia itself, can be troublesome.

Dock Ellis, who is unable to be here today because he is among a group of athletes currently making a tour in Vietnam, is a trait carrier and has health problems associated with that fact, which he described in his recent appearance before the Public Health Subcommittee of the Senate Committee on the District of Columbia. And Willie Stargell can outline for you today some of the experiences of his 9-year-old daughter, Wendy, who also is a carrier of the sickle cell trait.

But it is the disease itself, one must remember, that is the killer. The problems of the trait carrier are mild in comparison to the agonizing pain and the terror that the sickle cell anemia victim endures. Consider then that this disease strikes an estimated one in every 200, or even one in every 500, black persons born in this country, and you begin to appreciate the magnitude of the sickle cell anemia problem in the United States.

Because the disease is inherited, its spread can be halted, and the tragedy is that we are only now beginning to launch an attack of significant scope against it. There certainly can be no justification for any further delay.

In urging the quickest possible affirmative action on the legislation that is before you today, we pledge ourselves, too, to provide all the possible help that we can give in seeing that this fight is won.

This is a "must game," so to speak—a contest calling for the all-out effort of all who are interested in seeing victory achieved.

The roles that we believe our foundation can play in this struggle, on both national and local levels, are fundraising, education, and program coordination. Among the athletes who have agreed to help in the Black Athletes Foundation's plan to open offices and to become involved in testing and screening programs in every major sports city in the United States are Roy Campanela in New York City; Ernie Banks in Chicago; Bob Gibson in St. Louis; Willie Davis in Los Angeles; Jim Wynn in Houston; Walter Johnson in Cleveland, and Hank Aaron in Atlanta. There will be many more.

Although our foundation is quite new, having been incorporated in late May of this year, already we have been involved in the coordination of a pilot program in Allegheny County—that is, the Pittsburgh, Pa. Metropolitan Area, where our major office is located. And our experience there leads us to certain conclusions which, we believe, are relevant to a national sickle cell prevention program.

The Black Athletes Foundation offers the following four recommendations:

- (1) The bill now before you appropriately places great emphasis on the screening and testing aspect of a prevention program. Because electrophoresis is clearly the best method for carrying sickle cell testing on the massive scale contemplated by the bill, it is our strong belief that the legislation should specify that this method of testing be used.

Unlike the sickle-dex test and the sickle-prep test, which cannot accurately distinguish between the trait carrier and the anemia victim, the electrophoretic test can make such a determination. Additionally, it can pick up other abnormalities of the hemoglobin, such as hemoglobin C.

Although a rather expensive laboratory procedure when isolated samples are involved, its cost is as inexpensive as any of the other

sickle cell tests when large numbers of samples are to be run. In such cases, the total cost of the electrophoretic test, including collection of the blood and salaries paid to the technicians involved, approximates 50 cents per sample. The sickle-dex test, in contrast, costs about \$1.25 to \$1.50 per sample, whether small or large numbers of samples are to be tested. Factors of both cost and effectiveness, therefore, dictate the electrophoretic method in large-scale testing programs for sickle cell anemia.

(2) An important part of an effective attack on sickle cell anemia must be a strong followup plan not only for counseling trait carriers, but also for advising and treating those persons who have the disease. In the coordinated program in which we are involved in Pittsburgh, the Allegheny County Health Department, the federally funded Homewood-Brushton Health Center, the Central Blood Bank of Pittsburgh, the Bidwell Cultural and Training Center, the local Pittsburgh Sickle Cell Society, Planned Parenthood, and the University of Pittsburgh all are making contributions to this important followup phase to the screening and testing program.

Field interviews conducted by skilled personnel with trait carriers and anemia victims alike are essential to the program's success.

The identification of trait carriers provides such persons with the information that allows them, for the first time, to make crucial choices—in the selection of a marriage partner or in adopting children as an alternative to having their own. These choices clearly must be voluntary, as is, in fact, the decision to be tested in the first place. Yet, it is important that trait carriers be given the facts, impartially and without causing unnecessary alarm, about the risks they may face. While the very personal decisions about their families are their own to make, skilled counseling is crucial to stemming the disease in an atmosphere free from undue anxiety.

Nor can the sickle cell anemia victims be forgotten. As I mentioned earlier, there are probably about 50,000 such victims in the United States today. While there is currently no cure for the disease, its victims must be provided with the best possible treatment which may ease their suffering and prolong their lives. Many such patients will be from families whose levels of income are insufficient to meet the brutal financial demands that adequate care calls for.

Strong followup programs under the National Sickle Cell Anemia Prevention Act must be designed to meet both the problems of the trait carriers and the victims of the disease.

(3) It is our best estimate that the cost of properly doing the needed testing, screening, and followup work is Allegheny County, which has a black population of about one-quarter million, will run to about \$1.9 million. As a general estimate we feel that the cost of such a program will be between \$1.5 and \$2 million for every 250,000 blacks in the population. On that basis, we estimated, in our testimony on the bill calling for a demonstration program in Washington, D.C., that the program's cost would run between \$3,444,000 and \$4,592,000.

Yet, while Washington, D.C., is the city which has the highest proportion of blacks in its population of any city in the country, there are five American cities that have larger black populations than Washington's 574,000. They are New York City, with 1,500,000 black residents; Chicago, with 1,150,000; Detroit, with 800,000; and Los Angeles and Philadelphia, with about 700,000 each.

Clearly, these five cities are natural locations for the establishment of sickle cell centers described in section 1102 of title XI, under the proposed bill. It is our belief, however, that the \$5 million in each of the next 3 fiscal years, which the bill calls for in demonstration grants, is insufficient to properly do the job.

If our estimates on a Washington program are correct, the establishment of centers for these five additional cities alone will cost a great deal more. Yet, hopefully, demonstration grants also will be available for the implementation of other innovative suggestions as to how screening and testing can be accomplished in other regions. Large black populations of workers at plants that are not located in major cities, for example, might be reached by testing programs carried out at the jobsite. There are also the mobiles that can be moved from site to site. The legislation not only must provide enough latitude to allow innovative programs of this nature to be tried, but there must also be sufficient demonstration grant funds available to make this possible.

The problem of sickle cell anemia has been too long neglected. It is extremely important that, in now focusing public attention on this problem and in raising the hopes of so many that the problem will be met, sufficient funds be allocated to do that job.

It is our strong recommendation that the amount to be set aside for demonstration grants be doubled—to \$10 million in each of the next 3 fiscal years.

(4) Finally, we wish to emphasize again that the problem of sickle cell anemia differs from many other health problems in that more than just scientific expertise must be brought to bear. It is an educational proposition, too, in very large measure, as the proposed bill, with its major thrust in the area of screening and testing, makes very clear. For this reason, we believe, it is important that sickle cell programs receive input from those who are most affected—the lay person, as well as the professional, and certainly the black, whom this disease attacks almost exclusively.

The National Advisory Committee, that will coordinate the implementation of the sickle cell anemia program through the National Institutes of Health, sets a fine example of black involvement. It is essential to the program's success, however, that this example be followed up at the local level. Screening and testing is a program that will take place in the black communities of America, with its success dependent upon the widespread participation by the residents of those communities. Their confidence and participation can best be assured, we believe, if qualified black community leaders and black professionals play leadership roles in the planning and implementation of the program.

The Black Athletes Foundation, at the same time, is firmly of the conviction that the help of all Americans is needed to see that this fight is won. Our own board of directors is an integrated body, as is our medical advisory committee. The fundraising activities in which we've been engaged are directed to members of all races. And today we call upon all Members of the Congress to join the struggle by acting promptly and affirmatively on the bill you have before you.

There is, as we've tried to stress before, a great need for speed. A loss of any more time means a further loss of life. All of us, together, must help prevent that from happening.

Thank you, Mr. Chairman and members of the subcommittee. We would be happy to respond to any questions that you may have.

Mr. ROGERS. Thank you very much, Mr. Johnson, for a very fine statement. Mr. Symington?

Mr. SYMINGTON. Thank you very much, Mr. Chairman.

I have no questions at this time. It is an excellent statement, Mr. Johnson. Before you arrived, we put a good many questions to some of the doctors and public health officials which have given us a better understanding.

Your testimony has assisted in filling out the scope of our appreciation of this dread disease.

Mr. JOHNSON. Let me add a little more to this. In 1970, there was just \$100,000 spent on sickle cell research, and this is terrible when we consider that \$1.6 billion was spent for research in the United States.

You can see how anemic this is when you think of \$100,000 spent for research for disease that handicaps and attacks 23 million citizens in the United States; for example, \$4 million was raised in 1970 for cystic fibrosis, a disease that attacks one out of 3,000, 98 percent of whom are white; muscular dystrophy, estimated to affect one out of every 5,000 persons, received \$10 million; cerebral palsy had \$14 million for research last year.

Mr. ROGERS. Dr. Carter?

Mr. CARTER. Thank you, Mr. Chairman.

Certainly I want to compliment the distinguished gentleman for being here. It is a treat for me to see people I have watched on television performing magnificently. I am glad that you are bringing this to the attention of the American public. That is one thing that has not been done. This you are doing today.

For my part and I am sure the part of the committee, we mean to be helpful in this area.

Thank you, Mr. Chairman.

Mr. ROGERS. Mr. Kyros?

Mr. KYROS. Thank you, Mr. Chairman.

One thing in your testimony which I feel is important is the enlightened attitude toward the disease, and the importance of trying to advise everybody, so that marital partners who both have the trait would know the problems they face. The communities must be involved and the people in them won.

On page 7, you point out very accurately these are not easy decisions to make. I think groups such as yours who have a wonderful relationship with many black people in the United States will be very helpful.

You not only have to have the medicine, money, and research, but also the confidence of the people when dealing with them in an intimate way. I am delighted that you made that point in your testimony.

Mr. DAVIS. May I comment? You were saying something about the trait carriers, partners in marriage. We feel very strongly, unlike our county programs that we have with all the other health related agencies working in the art of screening and testing, that every county health department should have a computer blood bank. Now this computer blood bank is a standard procedure in most counties for persons to be tested for venereal disease. When two black individuals

apply for a marriage license, the computer blood bank automatically gives the blood sampling crew at the health department a feedback. If these two people have the trait then this is where the planned parenthood, which is part of our organization, immediately starts counseling.

If their names are not housed in the computer blood bank, then the county health department, which has a phoresis machine on its premises, automatically tests for sickle cell anemia. I listened to testimony prior to this. They were talking about establishing 20 clinics with \$1 million. Now it is the foundations feeling if people are going to set up 20 clinics and only give \$1 million, then we will go in another direction because I am quite sure we can raise a million dollars, \$1 million for 20 clinics, what are they accomplishing? What do you hope to accomplish? Here you are putting 20 clinics with \$1 million?

Mr. KYROS. That is \$50,000 per clinic.

Mr. DAVIS. Right. Something else that is very important. We are talking about metropolitan areas. Please do not forget about the people in Louisiana, Mississippi, Alabama. If we are going to make provisions for sickle cell anemia, let us include them because the largest part of our population is still in the South. We are talking about these centers here in these major areas.

Let us make some provisions for the people who are still in the South because they have the problems. In fact it is worse down there because the medical facilities are much more inadequate than they are in the North. So, in any legislation or bills pending, make sure the people in the South are covered by this legislation.

Mr. CARTER. Mr. Chairman, you will remember I brought out that very point. I would like to have it through the different health departments in our country, throughout the country.

Mr. KYROS. Thank you, Mr. Chairman.

Mr. ROGERS. Mr. Symington?

Mr. SYMINGTON. I was going to ask the question I put earlier to some of the other witnesses that really this testing should occur upstream, should it not, before they are getting ready to be married, but when still children perhaps, at least at the point when it is no longer masked. When once the sickle cell is detectable it ought to be detected.

When dealing with children I am always concerned about the voluntary aspect of the test. Mr. Johnson has mentioned that the decision to be tested should be voluntary. This ought to be examined in the sense that society has a voluntary opportunity to protect itself and perhaps through reasonable arrangements and agreements among parents and health officials that we can learn whether a certain condition exists in children. That is the only way we can provide properly for their medical attention.

Mr. DAVIS. I heard before that people were sometimes concerned about whether people will participate in a mass screening program.

In Pittsburgh we had a 3-day affair on mass screening where we actually had 5,000 people attending the screening. Of course we ran out of supplies and equipment and what not because it was a much larger crowd than we anticipated. We tested 3,700 people. We uncovered 300 people who were not knowledgeable they had the trait. Eleven had the disease itself.

The reason I am bringing that up is that there are some people who say it is very difficult to get people out to attend a mass screening and testing. I think if it is promoted properly that there is no question it has to be successful because in Pittsburgh our time was from 12 until 6 in the evening, yet at 9 'clock in the evening people were still lined up on the streets waiting to be tested for sickle cell anemia. I don't think there should ever be a question about where people are saying, "Well, it is very difficult to get people out for screening and testing." Right now people are waiting in Pittsburgh to be screened and tested.

The only problem we have is no money. I think this is the problem in every city today. So I feel if you act tomorrow on this, it is not too soon.

Mr. SYMINGTON. Thank you, Mr. Chairman.

Mr. ROGERS. I understand your feeling. Of course the money would be insufficient as proposed by some of the testimony today.

Now, what about the money proposed in the bill, is that a sufficient amount to do the job?

Mr. DAVIS. No.

Mr. ROGERS. What is your projection of a figure that is necessary now?

Mr. DAVIS. We have felt that for every 250,000 blacks, there should be appropriated anywhere from \$1.5 million. Now \$90 million in this bill, this is taking research, testing, screening, counseling, what not, that is over a 3-year period. It should be 4 times that amount. If they are going to appropriate billions for health research and you are going to give, let us say, a token thing to the blacks in the Nation where we are having a disease that affects 1 out of 10, you are talking about giving us \$90 million. I should not use the term that we should have our fair share. The disease deserves more than \$90 million a 3-year period. It would sound better at \$99 million every year.

Mr. ROGERS. I understand that you feel, too, that it would be important to have the blacks involved in the program, particularly, I presume, in fieldworkers, to get people involved and be willing to come in for screening and followup counseling?

Mr. DAVIS. Yes. This is important. The blacks do have some position in this disease because it is their disease in a way.

On the other hand, the blacks have demonstrated, especially the medical students from the University of Pittsburgh, the medical students from the other schools, that they are concerned and so I think quite definitely blacks should be included in the program.

Mr. ROGERS. Mr. Aaron, what do you feel the Black Athlete Foundation can do with the passage of this bill that it could not do now?

Mr. AARON. I think we would like to expand it, they are not doing anything in the State of Georgia on sickle cell anemia. I would like to get some of the people to come out and be tested. This is a very important point that the black athletes are making, contributions and going out and getting these people to come out and be tested.

I, for one, would like to see this expanded all over the United States, particularly Atlanta, Ga., because I am from that area.

Mr. ROGERS. I think that your foundation could do a great deal to promote interest in this problem and to encourage people to come in for screening as you have done already in some areas.

Dr. Georgio, what is your connection with the Black Athletes Foundation? You may have some feelings on this funding situation, too.

Dr. GEORGIO. I am in the Department of Medicine at the University of Pittsburgh, on full-time teaching and the research staff of the section on hematology. I have a clinical as well as research interest in sickle cell anemia and allied diseases for some time. I am also privileged to be a member of the Black Athletes Foundation and also on their medical advisory board.

I will address myself to the proposed funding. Just off the cuff arithmetic, if one screens 10 million black people in the United States in a year—it may not be possible to do that in a year—but just for the sake of arithmetic, one will find between 20,000 and 30,000 people in that 10 million with sickle cell anemia. If you then take the average hospital cost for a person with sickle cell anemia to range between \$2,000 and \$6,000 a year including the cost of blood and so forth and primary care, considerations are to be written in the bill, as I understand they are, and I think they should be, one can see that the fallout from screening will be immediately consumed just in terms of hospital care costs for those detected cases.

So I would agree with the gentleman from the Black Athletes Foundation that, indeed, the sum is not adequate although I would say it is certainly a good start but it must be expanded.

Mr. ROGERS. Thank you very much.

The committee is grateful to all of you for being here. Your testimony, I am sure, will be most helpful in helping the committee to come to some decision on the provisions of the legislation. We are hopeful that it will be reported out.

Thank you very much.

The next witness is Mrs. Harriet Karuhije.

We welcome you to the committee, Mrs. Karuhije.

**STATEMENT OF MRS. HARRIET KARUHIJE, ON BEHALF OF THE
AMERICAN NURSES' ASSOCIATION; ACCOMPANIED BY SUSAN
MOON, ASSISTANT DIRECTOR, GOVERNMENT RELATIONS DE-
PARTMENT**

Mrs. KARUHIJE. Thank you, Mr. Chairman.

I am Harriet Karuhije, a professional nurse, formerly an instructor of medical-surgical nursing and presently a doctoral candidate in nursing education at Teachers College, Columbia University, in New York City. Accompanying me today is Susan Moon, assistant director of the Government Relations Department of the American Nurses' Association. I am here today, representing the American Nurses' Association, to speak in support of programs for the prevention, diagnosis, and treatment of sickle cell anemia.

One of the major reasons for the existence of the American Nurses' Association is the promotion of optimum health care for all people. A large portion of our people, particularly black people, cannot achieve optimum health because of sickle cell anemia. It is estimated that 600,000 black Americans have sickle cell anemia and approximately 2.5 million black Americans have sickle cell trait. Although the sickle shaped red blood cell was first identified and reported in 1910, there

has been little interest in this health problem until very recently. Advances in science and technology must be directed toward the prevention, diagnosis, and treatment of this condition in order to reach our goal of optimum health for all people.

The American Nurses' Association stands for health care as a right unrestricted by considerations of nationality, race, creed, sex, age or socioeconomic status. Health care in this Nation is not universally available. Our poor and our minorities do not even have access to the less than adequate health care available to the more affluent.

Black people comprise a disproportionately large segment of the lowest income level in this society. Nurses, particularly public nurses, have been aware of the marked effect of sickle cell anemia and sickle cell trait on the individual's ability to cope with the normal stresses of living. To cite a vivid example, the common cold, a frequently health problem to children living in substandard housing, can be the terminal even for the child with sickle cell anemia. Certainly sickle cell anemia is only one of the many, many problems of Black Americans. Yet, this could be one of the problems solved somewhat more easily if funds are made available. It is a responsibility of Government to provide these funds.

Sickle cell anemia is one of the disorders of the blood routinely discussed in schools of nursing. The pathology as discussed in a classroom is often not very impressive until the student cares for the child of three who limps and has difficulty walking because of a blood clot to the brain developed during a crisis, or the 8-year-old hospitalized three and four times during the school year because of the painful crisis and the 20-year-old undergoing skin grafts in an attempt to heal large leg ulcers. Only at that time is the individual tragedy of the illness recognized.

Being "left back" in school, being fired from a job, being unable to support a family, being unable to "mother" children, dying at age 20 are problems quickly identified by students as agonizing and critical issues for the individual, family, and society. The cost of chronic illness to individuals, families, and communities, financially and emotionally, cannot be estimated. The cost is more than monetary, the cost is the loss of the individual's potential contribution to the society.

The student, particularly the black nursing student, asks why more hasn't been accomplished in the treatment of this illness when science and technology have made it possible to transplant kidneys, place artificial valves in hearts, and cure some cancers. What is the answer?

In the past decade, health care professionals and the American public have witnessed dramatic breakthroughs in medical care. The fact that the individual who has a coronary has a greater chance of survival today than 10 or 15 years ago is due to the efforts of the health care team. This same health team effort is needed for the fight against sickle cell anemia.

We support the establishment of a National Institute for Sickle Cell Anemia and the proposed functions of the institute. We stress the importance of grants, contracts, and traineeships being available to all members of the health team to upgrade their knowledge and skill in order to provide better health care services to the individual, the family and the community. Prevention and early recognition of illness is a primary health care role of nurses and other community health care

workers. Identification of the research and health care needs to combat this multifaceted disease requires the collaborative efforts of all providers of health care services.

Each members of the health care team should be included on the Advisory Council. Nurses must be members of the Advisory Council, they are vital links between medical therapy and patient services. The nurse has traditionally been the facilitator for individual and family entrance into the health care system and nurses are currently expanding this role to become the patient's advocate.

The major thrust of nursing activity will be directed toward initiating, planning, implementing and evaluating patient centers and health care in all settings.

The American Nurses' Association has since its origin 75 years ago recognized and worked for health as a national priority. The prevention, diagnosis and treatment of sickle cell anemia is another step in achieving this goal.

I thank this committee for the opportunity to appear here today. I will be glad to answer any questions that the committee might have.

Mr. SYMINGTON (presiding). Thank you very much. Mrs. Karuhije. I think your brief testimony is eloquent and important to this committee because it gives us another perspective of the problem encountered by people who suffer from this disease. A different kind of significant perspective.

I am wondering if there is a nurse on the Sickle Cell Disease Advisory Committee?

Mrs. KARUHIJE. I am glad you asked that question. The existing Sickle Cell Disease Advisory Committee is composed of representatives from social service, medicine and the community. There are no nurses included within the present structure. We feel very strongly that a nurse is a very important addition to the committee.

Mr. SYMINGTON. I anticipated that that might be our recommendation. I think we will certainly receive it sympathetically on this side.

I would like to know also how you feel about testing all children, particularly all black children, since this has been characterized as a disease that strikes the black race, testing if not children, at least young adults at some point during the growth pattern as a matter of course as distinct from waiting for symptoms.

Mrs. KARUHIJE. I see that question as one that would involve a great deal of ethical problems for one thing. That would be in a sense, should we make any health measure mandatory?

We get into the problem when we think about the mandatory acts for the different immunization programs. My own personal experience would lead me to comment that I think it should be a test for all children.

Mr. SYMINGTON. Thank you very much.

Mr. ROGERS. Dr. Carter?

Mr. CARTER. Certainly I have been impressed by your testimony. You raise a valid point about having a member on that advisory committee. I would personally recommend that.

Do you think that these screening tests should be mandatory for all black children in the country?

Mrs. KARUHIJE. That is related to the question that Mr. Symington just asked. I personally feel that they should be mandatory for if we are to have an effective screening program and we are going to

then use this screening as a basis for our genetic counseling, then certainly if we are going to allow some of the population to be not included, then we are going to be missing a major segment of the transferability of the condition if we don't test all of them.

Mr. CARTER. Do you think we should make this mass screening effort within the next year, that that should be part of our program?

Mrs. KARUHIJE. Personally, I think that would be an excellent suggestion.

Mr. CARTER. What do you think about screening people who intend to marry when their blood tests are taken as is necessary in almost every State now that I know of.

Mrs. KARUHIJE. I would answer that in two ways. If we were to do a mass screening program on all children, then eventually we would not need to do it on the marriageable age people.

Mr. CARTER. It might be a little too late, is that right?

Mrs. KARUHIJE. We would already have the information if we had an effective screening program of all children. However, I think that at the present time we need a concomitant type of approach. That would be the double approach.

Mr. CARTER. When the time comes for marriage and the bride and proposed partner have a genetic defect such as is characteristic, such as goes with sickle-cell anemia, it might be rather embarrassing and rather difficult, might it not?

What do you think of screening all youngsters when they are born, taking cord blood?

Mrs. KARUHIJE. There is a difficulty, it is a little too early to tell at this age whether the child has sickle cell anemia or not. It is only at the present time detectable at about 1 month. At birth it would not be a very effective test.

Mr. CARTER. I heard testimony to that effect a while ago but I believe that might be debatable. It is quite usual to take samples of cord blood in many of the hospitals and at the same time we do our urinalysis to check for phenylketonuria, PKU, and I think perhaps a test can be evolved which would show that.

You have made a very good statement. Certainly I support legislation which will be effective in this field.

Thank you, Mr. Chairman.

Mr. ROGERS. Thank you, Dr. Carter.

In this whole program I presume nurses could play a most effective role could they not?

Mrs. KARUHIJE. Yes, they could.

Mr. ROGERS. It is not necessary to have the doctor to do the screening and testing. The nursing profession could be most effective.

Mrs. KARUHIJE. I was going to comment on your statement. The nurse with her wide and continued contact with the population is both valuable and competent in case finding. She is already acquainted with the prevention, cause and treatment of sickle cell anemia. Her skills could be readily utilized in any program for prevention.

Mrs. ROGERS. Thank you very much.

Mrs. KARUHIJE. Miss Moon would like to make a comment about a discussion that we have had on some of the semantics or terminology that is inherent in the discussion of sickle cell anemia.

Miss MOON. Mr. Chairman, Mr. Karuhije alluded to some stigma attached to identification of yourself as being a sickle cell trait carrier or of having sickle cell anemia. Rather than use the term "disease," we should try to use other terms because, again of the stigma attached to this term, "disease."

Mrs. Karuhije agrees that it was very important to try not to identify it as a "disease" process, even though it might be a multifaceted problem. We wanted to get this in the record.

Mr. ROGERS. Thank you. We appreciate your presence here. Your testimony will be most helpful.

Mrs. KARUHIFE. Thank you.

Mr. ROGERS. Our last witness today is Dr. James R. Kimmey, executive director of the American Public Health Association.

STATEMENT OF DR. JAMES R. KIMMEY, EXECUTIVE DIRECTOR, AMERICAN PUBLIC HEALTH ASSOCIATION

Dr. KIMMEY. If I may, I will submit my prepared statement for the record and make a couple of comments.

Mr. ROGERS. Without objection your full statement will be made part of the record.

(Dr. Kimmey's prepared statement follows:)

STATEMENT OF JAMES R. KIMMEY, M.D., M.P.H., EXECUTIVE DIRECTOR, AMERICAN PUBLIC HEALTH ASSOCIATION

Mr. Chairman and members of the committee, I am Dr. James R. Kimmey, Executive Director of the American Public Health Association. Our association, which with its affiliated organizations comprises more than 50,000 individuals interested in the health of the public, is pleased to have this opportunity to comment upon the health problem represented by Sickle Cell Anemia. The emerging recognition of the importance of this disease as the cause of disability and mortality among black Americans is overdue. An important part of this recognition is represented in the legislative proposals currently under consideration by this Committee. As an organization with a commitment to prevention of disease wherever possible and to securing the highest level of health for the people of the country, the American Public Health Association shares the growing concern on the part of legislators, community leaders, and citizen groups that more needs to be done to bring relief to the individual suffering from this disease.

THE DISEASE

Sickle Cell Anemia is a hereditary disease characterized by an abnormal type of hemoglobin—the oxygen-carrying pigment in the red blood cell—which causes the cell's shape to be distorted under certain conditions. Patients with full blown cases of the disease suffer a variety of exceedingly painful and destructive lesions. The effects are produced through impairment in the blood's function of carrying oxygen to the tissues and through mechanical blockage of smaller blood vessels by collections of the abnormally-shaped red blood cells. The cells are in abnormal configuration only at times, and cause the afflicted individual to have crises characterized by severe pain, fever, weakness, and jaundice. In addition to the acute crises, patients with sickle anemia develop joint changes, chronic skin ulcers, enlargement of the heart, and lesions in other organs of the body. Although each individual patient experiences a somewhat different course of the disease, the course is invariably downhill and the majority of afflicted individuals die before reaching age 30. Moreover, this is not the sudden death of the coronary thrombosis or cerebral vascular accident but a protracted painful torture with a predictable fatal result.

SICKLE TRAIT

For each individual who suffers from the painfully progressing and ultimately fatal sickle anemia, there are ten who have sickle cell traits. Generally, this is a relatively harmless condition and the individual remains undetected unless some unusual stress causes a crisis. Since the individual sickle cell trait is a genetic abnormality, it is possible for him to pass on the trait to his offspring. Thus, individuals with sickle cell trait are *carriers* and contribute to the retention of the hemoglobin abnormality in the genetic pool for generations.

GENETIC ASPECTS

Sickle cell anemia falls in a general category of disease known as *hemoglobinopathies*. The carriers of this disease have both normal hemoglobin and an abnormal hemoglobin called *hemoglobin-S* in each of their red blood cells with the normal hemoglobin predominating. If two carriers mate and both contribute the abnormal gene to the offspring, the offspring will be *homozygous*—that is he will have hemoglobin-S in his red blood cells and will develop sickle cell anemia. It is widely thought that hemoglobin-S is a mutation that is developed in Africa which has a tendency to protect infants against the prevalent and highly dangerous form of malaria in that part of the world. In the United States, this African origin of the disease has made it primarily a disease in Americans of African descent.

It is estimated that from 7 percent to 14 percent of blacks in America have sickle cell trait. The disease of sickle cell anemia occurs with less frequency with incidence varying from 0.3 to 1.3 percent.

NEEDED PROGRAMS

Considering the number of Americans afflicted by this disease, a greater investment in research and particularly in programs directed towards preventing the occurrence of full blown sickle cell anemia is fully justified. Earlier this week, the HEW Sickle Cell Disease Advisory Committee recommended expanded efforts in both of these areas to Secretary Richardson. Their recommendations included comprehensive centers of research and service, screening and education clinics, collaborative research and development, biomedical research, training of health personnel, and a program of public and professional information. In addition, the bills currently before this Committee—H.R. 11171 and H.R. 7654 deal with many of the same areas.

These administrative and legislative recommendations reflect a heightened concern for the importance of this disease in a significant segment of our population. It would seem important, however, that to the maximum possible extent programs for individuals with sickle cell disease be conducted as a part of ongoing health programs at the Federal, State, and community level. For example, screening tests for sickle cell disease should become a routine part of the examination of school age children at the time of their admission to school and near the end of their elementary school experience. Similarly, testing for sickle cell disease should be included as a routine part of the work-up of patients at risk in clinics, emergency rooms, and at the time of admission to a hospital. It is important that screening for sickle cell hemoglobin be incorporated in premarital testing for the population at risk. Since genetic counseling of carriers is the major means available for preventing occurrence of the disease, carrier identification before marriage is of particular significance. Finally, the specialized laboratory techniques for detection of the abnormal hemoglobin of sickle cell anemia should be made available throughout the country.

PROGRAM ELEMENTS

Research and Research Training

The need for basic research and research training in sickle disease requires that these subjects be given special attention. Our basic knowledge concerning effective treatment of the disease, once manifest, is slim and needs development. In addition, further research effort is needed regarding diagnosis and prevention. Although research goals in sickle cell anemia may be met without creation of a special governmental focus for such research, encouragement should be given to development of research centers at the state and community level where such studies could be undertaken.

Community Programs

Given the current state of knowledge of this disease the most important efforts that could be undertaken at this point in time are those directed toward early detection of both afflicted individuals and carriers coupled with organized treatment and genetic counseling programs. Since this type of activity must be made a part of ongoing preventive and therapeutic programs, special attention should be given to projects that encourage incorporation of sickle cell hemoglobin testing in school health program, and the programs in neighborhood centers and public health clinics, as well as other sites where individuals at risk routinely contact the health system such as hospital emergency rooms, outpatient clinics, and admission departments. Furthermore, laboratory procedures that are currently well-developed and understood should be incorporated into the spectrum of tests offered by state and local public laboratories as a service to individual practitioners in their areas.

Public Education

Support should be made available for programs aimed toward wider public understanding of the nature of this crippling disease and of the importance of detection of the carrier stage if future generations are to be protected. This program of public education should go hand in hand with those directed towards increasing the availability of screening tests and the necessary genetic counseling.

SUMMARY

Sickle cell anemia represents a serious killer andcrippler that has received relatively little attention compared to other conditions of less significance to the population. The current interest in this disease represents an opportunity to redress the historical imbalance. It is important that expanded research and detection programs be instituted, and such programs be integrated to the maximum possible extent into routine health programs for the population at risk. Since the disease affects mainly black Americans, and since this group of the population are denied access to medical care for a variety of social economic reasons, special attention is needed in the form of additional targeted detection, treatment, and counseling programs which would reach this population group specifically. Among the access points that should be covered in such programs are the public schools, hospital clinics and emergency rooms, premarital exams, and armed forces induction procedures.

Dr. KIMMEY. The sickle cell process has been adequately described here both in terms of the underlying pathology and the manifestations in patients. It has been described only in the abstract although some mention has been made of the type of things that happened to people from this disease. I think it is important when you talk about a disease that affects literally millions of people to remember that in the final analysis the disease affects one individual at a time and these are very miserable, unhappy, sick people.

I am sure Dr. Carter has seen patients with the terminal phases of this disease and it is one of the ugliest and most unpleasant ways to die that I have seen in my experience.

I think there has been a general agreement among the witnesses and among the bills before you as to the scope and importance of this problem. Yet, they all suggest different approaches and levels of effort. I would like to comment on these matters.

I think the first priority as we are beginning to take long overdue action in the matter of sickle cell anemia should be in the area of community programs. Community programs of detection, counseling, of treatment of the cases that are found, and of organized followup. A number of the tests that are available for screening for sickle cell anemia have been described earlier.

One of the issues which has been raised several times is the matter of screening in schools as well as to premarital screening. I think it is good to screen at the time of school entry and also again at the end of the elementary school experience.

Premarital screening probably has considerable value in finding carriers but there are problems in premarital screening since your choices are limited. It takes skillful counseling to explain to a couple the importance and the impact of the disease and the possibilities that two partners have for producing affected offspring. The earlier the detection of the disease can be made, probably the better.

I might comment that there are many conditions for which we have routine tests. Routine tests are made when someone is brought into the emergency room. People brought into the hospital have a battery of tests. I think, considering the importance of this disease and the number of possible cases within the population, that our major move ought to be to have this incorporated into the routine practice of medicine for the population at risk as quickly as possible. The testing procedures are available. This is not, of course, to the exclusion of research.

At this point of time, if we are going to begin to screen for this disease, the place to plug it in is routine activity, in health clinics, emergency rooms, neighborhood health centers, and so forth. This is not a substitute for having some highly specialized centers for study of sickle cell disease but certainly a supplement we would recommend.

Mr. ROGERS. Why can't this be done right now with some urging of the Public Health Service? Don't you think this could be done very quickly?

Dr. KIMMEY. The State health officials will be in 2 weeks from now for their annual meeting with the Surgeon General. I don't know if sickle cell screening is on the agenda. It is a point that might be raised with them. Certainly in many States, particularly the States with high populations at risk, I see no reason why they could not begin to explore this actively.

I think they would turn back to the Congress and the Department and say, "Yes, but we will require funding," and of course that is what we are talking about today. We do have to have the funding for the programs. I don't think you would get much resistance on the part of the Public Health Departments to conducting the programs if the funds are available.

Mr. ROGERS. How much would the cost be to do the additional test on the blood before marriage?

Dr. KIMMEY. At this point in time they suggest that the cost—I think this is in some of the earlier testimony—is about 50 cents.

Mr. ROGERS. What about it?

Dr. KIMMEY. That is the screening type test. If you are going to premarital testing, that is a screening test. You have here the most important population to get to right now, people who will have offspring with the disease and 50 cents is certainly not too much.

Mr. SYMINGTON. What is a definitive test?

Dr. KIMMEY. Hemoglobin electrophoresis. This will tell whether one has the trait or actually has the disease.

Mr. SYMINGTON. You are speaking of definitive tests of that condition alone, not definitive tests for the number of possible conditions?

Dr. KIMMEY. I was talking about the hemoglobin. Electrophoresis

would pick up some of the other hemoglobin diseases, many of which are genetically inherited. You pick up more than just sickle cell. Of these diseases, sickle cell is the most common in this country.

Mr. SYMINGTON. We are trying to get the answer from HEW now and they don't seem to be able to find their file. I addressed a question to them as to which States require a Wasserman test. Those that do would seem to do so for a reason. That is for the so-called social diseases. I am not quite sure how to distinguish a social disease from any other kind when it infects children, or can. That is why we elicit your views as to whether or not tests of this kind should be across the spectrum of possibility so that people are forewarned and therefore forearmed, you might say, in their decision.

Dr. KIMMEY. It is vitally important if we get into the business of premarital examination that we make some investment in developing genetic counseling. Counselors need to sit down with the people detected and explain to them what has been found and what its implications are. The earlier testimony about the one in four business, I am sure that people, if this is not adequately explained would think it is one child in four when it is a one in four chance for every child. These are sophisticated concepts. In order to explain them adequately so that people can make informed decisions takes a certain amount of skill, training, and understanding. This is a concomitant investment along with the screening.

Mr. SYMINGTON. This one in four business, I would like to clarify that. I thought I understood that one in four could be expected to have the disease and two others would be expected to be a carrier. Only one in four could be expected to be free as far as the carrying characteristic or the disease?

Dr. KIMMEY. That is right, but these chances recur for each conception. You could have families in which the lightening has struck thrice and you have three children with the disease or three that are not even carriers or three that are carriers or any permutation thereof.

I will add one more thing.

The sickle cell anemia problem points up for us one of the defects in the way we establish priorities for investment in the health system, whether it is Federal investment, State investment, or investment of the individual physician's time. Here is a problem with a very high death rate in the population and for the black population specifically it is extremely high. Somehow it has not gotten the attention of the Department or Congress. PKU, which you have heard of before, affects something like one out of 18,000. Yet PKU is a disease for which many states have installed test requirements and subsequently stopped because of the low yield. The required screening test at the time of birth at the time I was delivering babies in Ohio, was to test the diaper for PKU.

We do have a priority problem. It is a mixed problem. It may have to do with lack of voluntary organizations. I think the black Athletes Foundation will help in this regard. You may have some overtones of the relative power and strength of the black community in relationship to national affairs.

I think it is high time we begin to turn priority attention to some of the things like sickle cell which is by and large identified with different racial characteristics.

Mr. ROGERS. Dr. Carter?

Mr. CARTER. I have no questions.

Mr. ROGERS. Mr. Symington.

Mr. SYMINGTON. There is some incidence of sickle cell anemia in whites, I take it?

Dr. KIMMEY. It is extremely low.

Mr. SYMINGTON. Is there any pattern there of who they are, what income groups, associations?

Dr. KIMMEY. Not that I am aware of. To see sickle cell anemia fully expressed in a white is exceedingly rare.

Mr. CARTER. That is why malaria is chronic among whites in the Mediterranean areas. It is thought that sickle cell anemia is sort of a mutation in response, a defensive mechanism.

Dr. KIMMEY. In this country it is very low.

Mr. SYMINGTON. That 1 percent figure was for North American whites who are not subject to malaria.

Mr. CARTER. They have their origins largely in the Mediterranean area.

Dr. KIMMEY. You would have to go back to look at family origins. It is, as pointed out earlier, a protective mutation.

Mr. ROGERS. Actually you were talking about this test earlier. Most of those children with PKU, if they have something not done then the general result would be retardation.

Dr. KIMMEY. That is right. PKU is different from sickle cell. If you detect it you can do something about it.

Mr. ROGERS. In that program even if you find two or three you will have paid for the whole program in that State by saving the cost to society of taking care of a retarded child.

Dr. KIMMEY. Quite so. If you can also prevent the birth and the first 20 years in sickle cell you have accomplished the same thing.

That is a natural consequence of prevention.

Mr. ROGERS. Thank you very much for your presence here today.

I might say we have two very distinguished gentlemen who are vitally interested in the public health field, who publish Family Child magazine—the chairman of the board, and Mr. White. We are honored to have you with us today.

This concludes our hearings. The committee stands adjourned.

(The following statement and letters were received for the record:)

STATEMENT OF HARLEM HOSPITAL CENTER, CALVIN H. SINNETTE, M.D., CHAIRMAN,
COMMITTEE ON SICKLE CELL AND ALLIED DISEASES

Harlem Hospital Center, in recognition of the serious problem posed by sickle cell disease on the black population and in recognition of the inadequate resources presently available to provide the full range of services for afflicted patients and their families, declares its wholehearted support for the efforts of Congressman Charles Rangel and other legislators to secure legislation with respect to this disease.

In particular we wish to endorse H.R. 11171 which seeks to provide grants for sickle cell screening and counseling programs. This aspect of service is sorely needed to provide correct information to the public and to develop an approach for preventing the disorder.

An indication of the community concern regarding sickle cell disease is the over 675 signatures we have obtained in support of the proposals of Congressman Rangel and other legislators which are appended to this statement for incorporation into the record on hearings for sickle cell legislation.

NATIONAL CONGRESS OF PARENTS AND TEACHERS,
Rock Island, Ill., November 13, 1971.

Re Sickle Cell Anemia

HON. PAUL G. ROGERS,
Chairman, Subcommittee on Public Health and Environment, House Committee on Interstate and Foreign Commerce, House Office Building.

DEAR REPRESENTATIVE ROGERS: On May 25, 1971, in convention assembled, the National PTA delegate body voted to support funds for research concerning sickle cell anemia.

The National PTA, with a membership of approximately 10 million, represents the interests of all the children of our great nation. Great concern was expressed for the magnitude of the problem of sickle cell anemia and the lack of research being done in this field. Since this is a disease primarily among the black people, and many of our members are black, this is a personal tragedy to them and their children, with little hope at the present in combatting the disease or bringing comfort to those afflicted.

We are pleased that legislation has been introduced to provide this much needed research. I was not aware of the hearing set for Friday Nov. 12, 1971 in time to ask that we be heard. Therefore, I would now hope that you would be willing to include our support in the findings of this committee. At this point in time when so much can be done and is being done to provide the greatest opportunity possible for all our children, we wholehearted support legislation for the prevention of sickle cell anemia.

Thank you for hearing our views.

Yours very truly,

Mrs. WALTER G. KIMMEL,
Coordinator of Legislative Activity.

HOUSE OF REPRESENTATIVES,
Washington, D.C., February 9, 1972.

HON. PAUL ROGERS,
Chairman, Subcommittee on Public Health, and Environment, Interstate and Foreign Commerce Committee, U.S. House of Representatives.

DEAR MR. CHAIRMAN: Enclosed is a copy of the resolution passed by the FDR-Woodrow Wilson Democrats, Inc., of New York City which directly relates to the hearings being held by your Subcommittee on sickle cell anemia.

I would appreciate your making this resolution part of the official records of the hearings for the consideration of the Subcommittee and the Congress.

With warm personal regards, I am,

Sincerely,

CHARLES B. RANGEL,
Member of Congress.

FDR-WOODROW WILSON DEMOCRATS, INC.,
New York, N.Y., January 31, 1972.

Representative CHARLES RANGEL,
House of Representatives,
Washington, D.C.

DEAR SIR: The following resolution was passed unanimously at the January 24 Board of Directors meeting of the FDR-Woodrow Wilson club:

"That we urge our Representatives to legislate funds for research on Sickle-Cell Anemia as an urgent health priority. Roughly 1 out of 500 blacks suffers from this disease—a suffering which is as often as not excruciatingly painful.

"We also urge our Representatives to legislate funds for the treatment of those who have the disease."

Sincerely Yours,

MICHAEL MERRYMAN
 (For the Board).

(Whereupon, at 12:45 p.m., the committee adjourned.)



