New Directions

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Sickle Cell Anemia: A Growing Interest in an Old Malady

Editorial Staff

A Growing Interest In an Old Malady

Sickle Cell Anemia

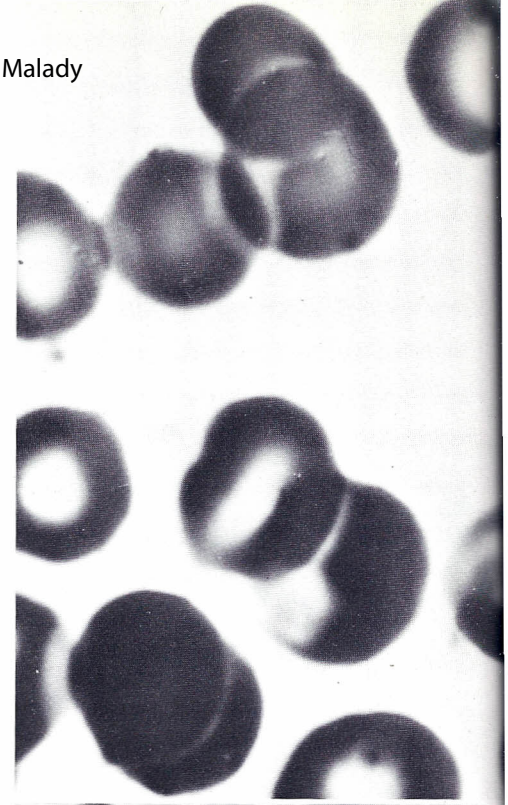
After many decades of neglect, coupled with indifference, by the Federal Government and private volunteer health organizations, sickle cell anemia—an inherited blood disorder that medical scientists say afflicts 1 out of every 500 Black Americans—has become a *cause célébre.*

Pressure from Blacks in high positions and increased awareness by the public about the nature of this illness are among the key factors responsible for this new development.

This newly-found interest in an old malady that was identified clinically 63 years ago—still with no effective cure—emerged a few years ago. It became a national cause following President Richard M. Nixon's February 1971 health message to Congress when he called sickle cell anemia a high priority target and asked for $5 million in Federal expenditures to fight the disease during fiscal 1972. Congress subsequently increased the request to $10 million. And the Secretary of the Department of Health, Education, and Welfare appointed 11 prominent individuals to serve on the Department's Sickle Cell Disease Advisory Committee, "to advise on objectives and priorities."

In his message, Mr. Nixon said sickle cell anemia was second only to cancer in the Government's program of national health priorities. But about the same time, the Government made a request to Congress for $1.6 billion for a three-year program to fight cancer. Critics, most of them Blacks, charged the Administration with double standards in dealing with problems of Blacks, and strongly asserted that the money set aside to fight sickle cell disease was not enough to wage an effective nationwide campaign against one of Black America's long-neglected health problems.

Ironically, instead of making adequate funds available to fight sickle cell, critics said the Nixon Administration at one point attempted to block a bill introduced by Senator John V. Tunney (D-California), to provide $142 million for a three-year campaign against the disease.



blood cells before any sickling occurs.

The Tunney bill passed the Senate unanimously in December 1971 but was later reduced to $105 million in the House. It was signed by the President in May, 1972 as the National Sickle Cell Anemia Control Act.

Although the Act authorized expenditures of $105 million in three years ($25 million for fiscal 1973 and the rest for fiscal 1974 and 1975), whether the full amount authorized by Congress would actually be appropriated is another question. In view of the Administration's budget-cutting attitude, the likelihood of substantial reduction in sickle cell funds cannot be precluded.

Black institutions like Howard University and other organizations engaged in sickle cell research and education programs are expected to receive more money when the Federal funds are made available. What portion they will get is unknown.

In the past two and three years, sickle cell anemia has become a popular cause for politicians, businessmen, entertainers and fund raisers.

"Sickle Cell Anemia Month" was proclaimed twice in Washington, D.C., by Mayor-Commissioner Walter E. Washington. Other cities have done similarly.

At least seven states and the District of Columbia passed measures requiring compulsory testing (for sickle cell trait) of Black schoolchildren. But unexpected opposition from doctors and community leaders who feared that such laws could hurt Blacks, has apparently caused the modification of some of the hastily adopted measures. The controversial District of Columbia measure, which would have required the compulsory testing of all Black schoolchildren beginning in September, has been modified. As a result, the testing of schoolchildren in the District will be done on voluntary basis.

Some months ago, Dr. James E. Bowman, director of laboratories at the University of Chicago and an outspoken critic of laws requiring compulsory testing for sickle cell trait, questioned the legality or morality of the District of Columbia measure (now revised) and a similar one in Virginia. He said, at a sickle cell symposium in Washington, D.C., he was against measures that would single out Blacks for genetic screening and warned that, if they went unchallenged, such measures could be used in a manner that is not in the best interest of Blacks in this country. A number of the 400 delegates attending the symposium agreed.

What is Sickle Cell Anemia?

Sickle cell anemia is an inherited blood condition, which for the most part occurs in a relatively benign carrier called sickle cell trait.

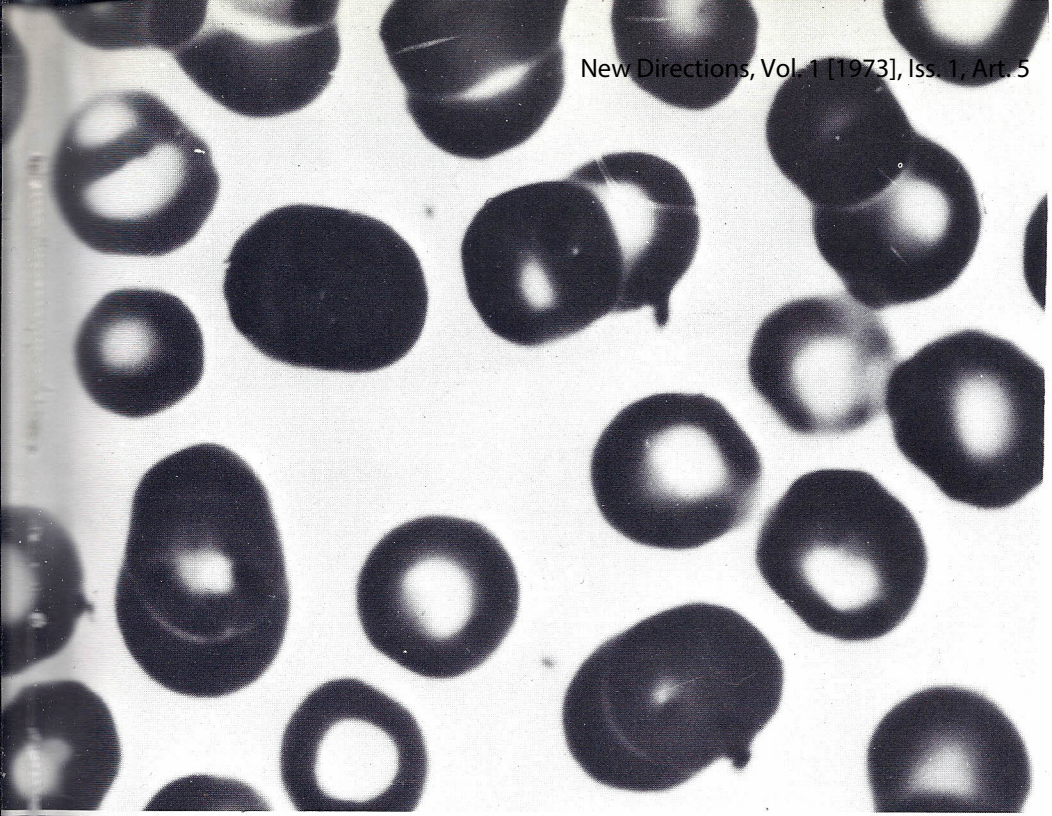
Approximately 1 out of every 10 Black Americans, or about 2.5 million of the Black population in this country, carry only the gene for the sickling trait, NOT THE DISEASE. Medical experts estimate the deadly form, the actual disease, attacks 1 out of every 500 Black babies born in this country. The number of Black persons suffering from the disease is estimated at 50,000 nationwide.

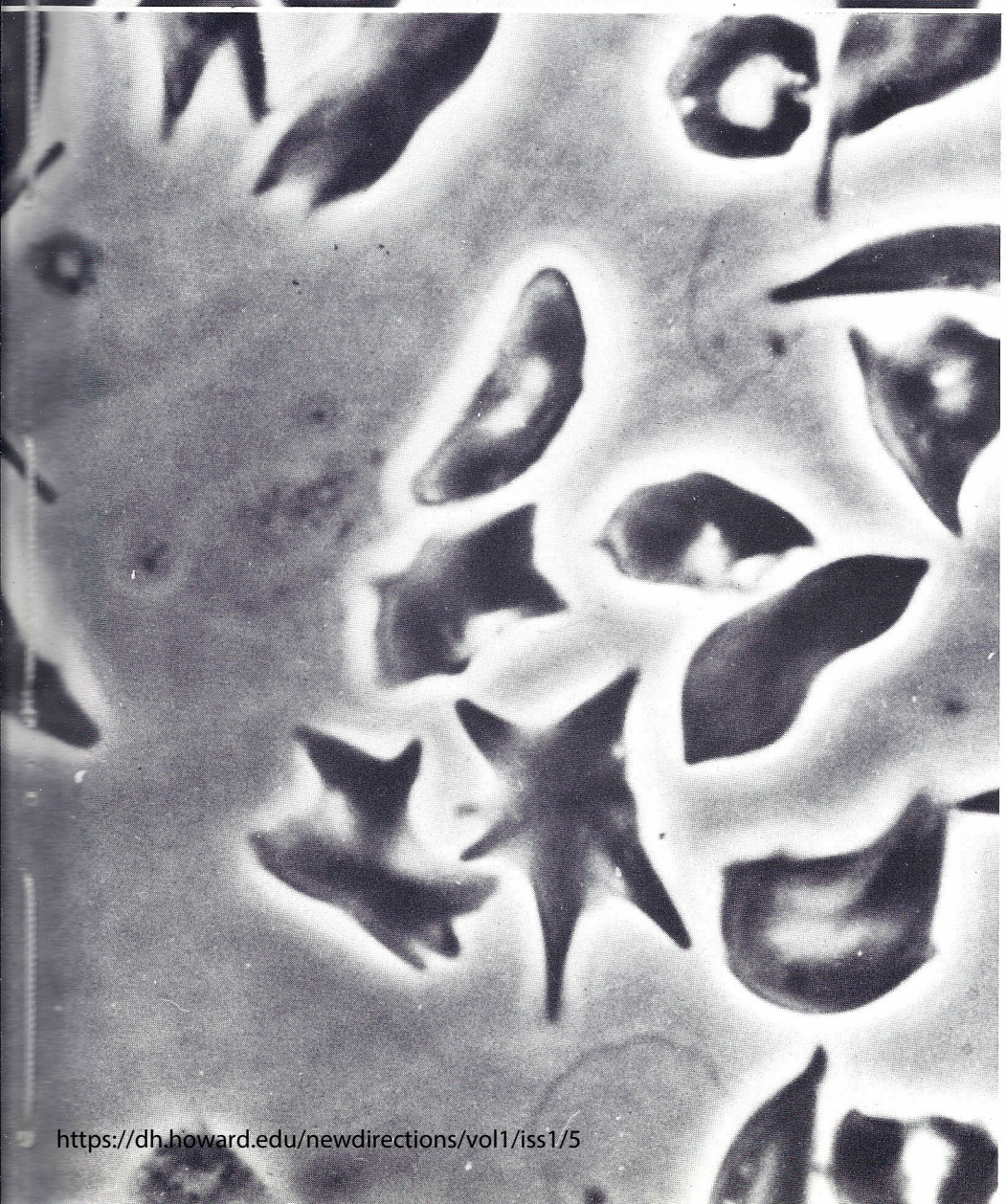
Sickle cell disease is caused by a genetically determined defect in hemoglobin, the respiratory pigment in red blood cells. Because of this defect, the oxygen supply throughout the body is diminished and the abnormal hemoglobin molecules stick to one another causing the normally doughnut-shaped red blood cells to sickle.

The sickling condition impairs the flow of blood to vital organs, such as the spleen, lung and the central nervous system, resulting in some damage to the organs.

Complications from the disease include susceptibility to respiratory infections, chronic anemia, recurrent symptoms of pain, headaches, swollen bellies and joints; and under extreme conditions, blindness, leg ulcers, crippling paralysis and convulsions.

The victims experience painful attacks several times a year. Medical experts call





Sickle-Shaped Red Blood Cells, abnormal cells, such as those seen in the photo below, not only give sickle cell disease its name but are also responsible for the crippling and lethal effects of this inherited blood disease.

the recurring painful episodes "crises."

Because of the recurrent attacks, children suffering from the disease are usually bedridden several days, sometimes weeks, during a school year. Frequent absence from class causes some children to lag in their studies. Also, young victims of the disease can not participate in most strenuous physical activities enjoyed by their healthy peers. This condition of limited childhood activity creates psychological problems that, in most instances, require expert counseling.

Adult victims suffer similarly, but to a lesser degree because they learn to cope with their handicap as they grow older. Sickle cell anemia is a physical illness. It does not affect the mind.

The disease does not affect all Black people either. It attacks only some offspring of parents carrying the trait. If two persons with the trait marry, their children would have a 25 percent chance of carrying the disease, 50 percent chance of carrying the trait, and 25 percent chance of being normal. That is—one out of four offspring would be born with the disease, two out of four would have the trait and one out of four would be born normal.

When either the father or the mother is afflicted with the disease, all offspring are born with the trait. When either the mother or the father carries the trait, the children have an even chance of being born normal or with the trait. In most instances only one parent is trait carrier.

Medical experts say persons carrying the trait do not usually show physical defects. In fact, they seldom have knowledge of it unless given a simple blood test.

All carriers of the trait, and victims of the disease, are born with the condition. In victims, the disease usually surfaces about the age of two, requiring costly medical attention the rest of their lives for those who live to the median age of 20. The fatality rate before the median age is

20 approximately 50 percent, according to medical experts.

There is no known cure at the present time. However, the discovery of antibiotics has contributed to the easing of pain and to controlling complications. Meanwhile, researchers continue to test new drugs. Doctors say good medical attention, along with proper home care, can make it possible for those afflicted with the disease to live comfortably most of the time.

Medical experts in this country trace sickle cell anemia to Africa, where they say the trait developed centuries ago as a bodily defense against certain types of malaria.

However, a medical scientist from West Africa told a different story when he addressed delegates at the symposium held in Washington. Dr. F. I. D. Konotey-Ahulu, a Ghanaian, said sickle cell's effectiveness as protection against certain types of malaria was for a limited time only, becoming neutralized when those born with the malady reached the ages of two or three. The protection against malaria did not help adults, he noted.

Dr. Konotey-Ahulu also explained that contrary to the general belief of some people in this country, sickle cell anemia is by no means an exclusive malady of Black people. He said its victims include Europeans who live in the area of the Mediterranean Sea, particularly Greeks and Italians.

Disease Identified 63 Years Ago

Sickle cell anemia was first identified clinically in Chicago in 1910. One of a handful of significant developments was achieved in 1949 when a chemist in California determined that an abnormal hemoglobin was responsible for the condition. In 1970, a team of doctors in Michigan announced a promising but still controversial therapy: Treatment by urea, an organic waste substance administered intravenously to halt pain temporarily. However, the drug is still being tested to determine its effectiveness. Also under test in New York is another recently developed drug called potassium cyanate.

The Future Holds Promise

In many cities across the country, programs to combat the disease have been developed during the past years. Various groups consisting of professionals, athletes, artists and lay persons have sprung up in several places with predominantly Black populations, such as Washington, Baltimore, Philadelphia, Pittsburgh, New York, Detroit and Los Angeles. Their objective: To raise funds or be involved in some way in a popular cause.

Suddenly, sickle cell anemia is now receiving national attention. The Government is giving it the attention it should have years ago but did not because, as Dr. Roland B. Scott says: "It (sickle cell anemia) occurs in Black people who have low priority in our culture."

A nationally recognized pioneer in sickle cell anemia research, Dr. Scott is Professor of Pediatrics at the Howard University College of Medicine and Director of the Center for Sickle Cell Anemia. Using facilities at Howard, Dr. Scott has done extensive study and research in sickle cell anemia for more than 20 years, often with meager financial support.

There is no known cure for sickle cell disease yet, although research efforts are continuing at various levels. But for research to continue at an intensified pace, more funds are needed.

The $10 million approved by Congress has enabled the Department of Health, Education, and Welfare (HEW) to expand existing programs and create new ones in the continuing effort to conquer the disease.

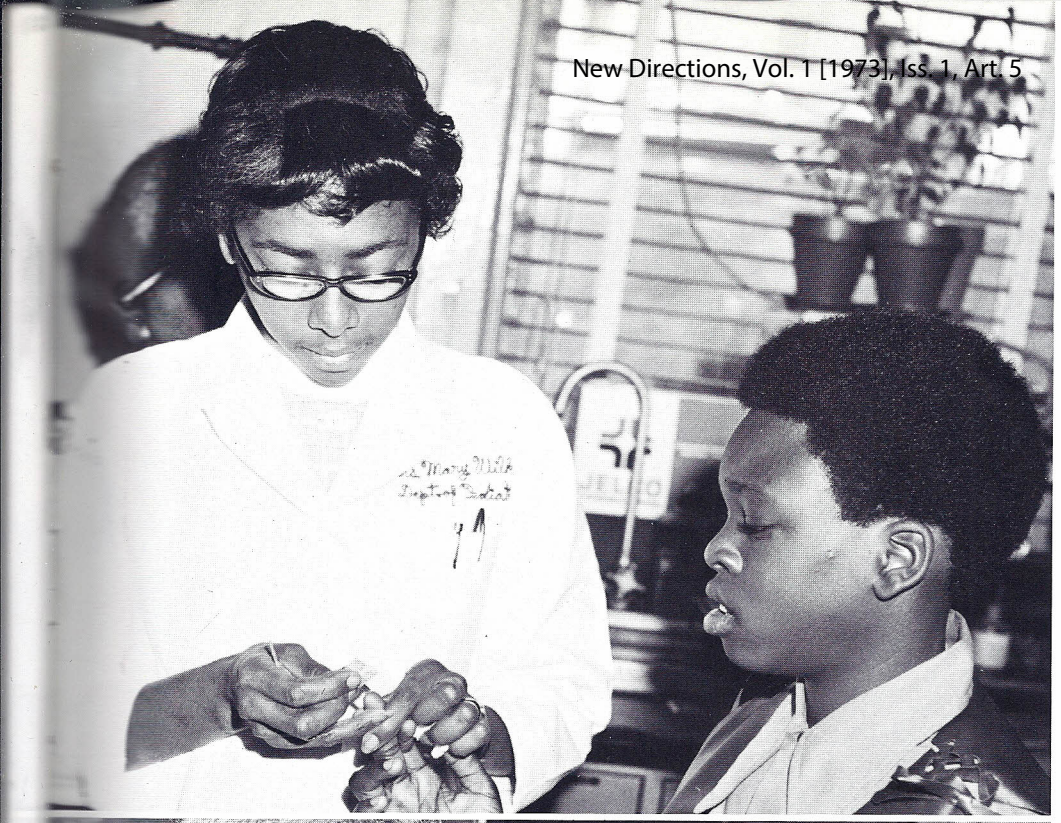
Despite poor response in the past, the Government is now showing signs of willingness to support the sickle cell anemia cause. Its revitalized efforts have drawn praise as well as criticism. Some Blacks say the Government is not doing enough and that more needs to be done. Other Blacks describe the new thrust by the Government as a hoax. Still, others say sickle cell anemia is not a major health problem of Blacks, nor it is as deadly as the public is led to believe. While the debate continues, some positive actions have occurred.

In March 1972, Dr. Rudolph E. Jackson, a Black physician from Memphis, Tenn. was appointed by the Secretary of HEW to head the Government's intensified program to fight sickle cell disease. Dr. Jackson is coordinator of the National Sickle Cell Disease Program under the National Heart and Lung Institute (NHLI), a unit of HEW's National Institutes of Health, (NIH). He also is head of NHLI's Sickle Cell Disease Branch.

In July 1972, HEW announced the awarding of grants and contracts worth $9 million to support 10 comprehensive Research and community service centers (including the Center for Sickle Cell Anemia at Howard); 27 new and 7 existing collaborative research and development programs dealing with the nature and treatment of sickle cell disease, and 19 screening and education clinics in Black communities for counseling, screening and definitive diagnosis of sickle cell anemia. The largest single HEW grant, $826,057, went to Dr. Scott and his colleagues at Howard University.

Earlier, the NHLI awarded six contracts of more than a half-million dollars to principal researchers at six institutions (including Meharry Medical College which received $99,398) to conduct studies on three forms of promising therapy. One grant of $179,028 went to Travenol Laboratories, Morton Grove, Ill., which in turn awarded a contract of $54,000 to Howard University, for the study of urea, a treatment already in use on an experimental basis.

In 1971, the Government spent close to $1 million on sickle cell anemia related research nationwide. Recipients, as noted by a spokesman for the HEW, were private and public institutions.



Mary A. Williams, a technician, takes blood from a young boy by pricking one of his fingers, a simple and painless process.

These expenditures do not impress many Blacks. For instance, soon after the government announced that it had allocated $5 million for sickle cell, Jerry Fowler, a staff member at a private sickle cell anemia program in New York, characterized the amount as "a drop in an empty bucket." Other Blacks charged tokenism.

Dr. Scott put it this way: "It is a small amount. We at Howard alone could use $5 million on research and related services, more space, additional staff, equipment and supplies."

Dr. Scott's views have not changed—even after the Government doubled its original $5 million expenditures for a nationwide campaign against the disease, and after almost one-tenth of it was awarded to Howard.

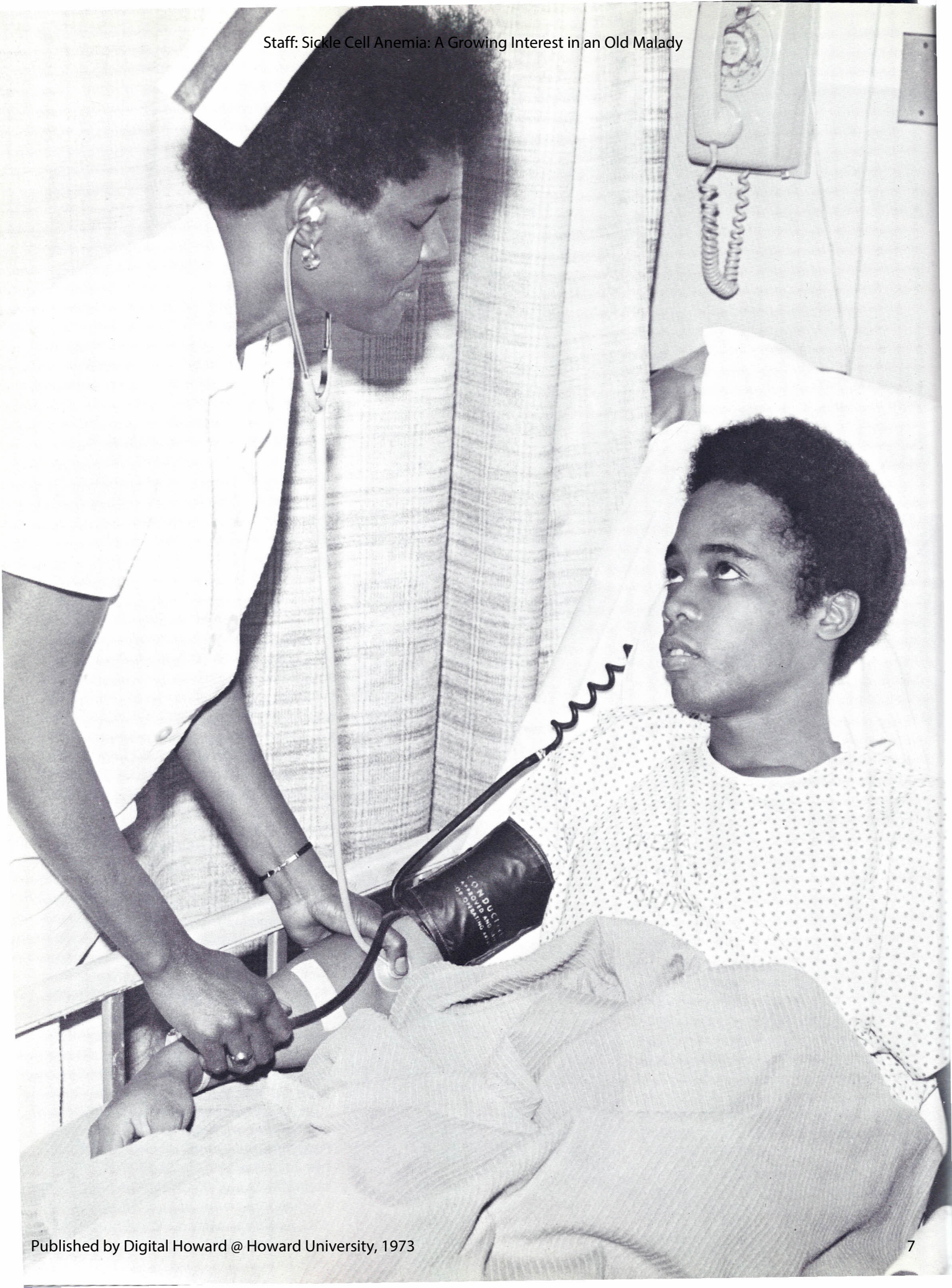
"When you are talking in terms of the whole country, $10 million is a very small amount," he said.

As for the $826,057 awarded to Howard, Dr. Scott had this to say: "We are grateful, but our needs have not been satisfied. It (the money) is going to help. The amount we got is 45 percent less than what we applied for. We still need additional funds."

For instance, during the 15 years prior to 1972, only $47,000 was received from the Government by Howard University for sickle cell anemia research, Dr. Scott said. Another $54,000 was received indirectly from Travenol for the study of urea.

During the same period, private organizations gave more than a half million dollars to Howard for sickle cell research, including a $273,000 grant by the John A. Hartford Foundation which was awarded to the College of Medicine for the study of "Clinical Aspects of Sickle Cell Anemia in the Newborn and Fetus." Dr. Verle E. Headings, Associate Professor of Pediatrics and Head of the project, said the objective is to find effective means for

Technicians like **James Ross** test blood samples to determine the presence of sickling traits.



Nurse **Cora Harvey** visits a patient to check his condition.

early diagnosis and treatment of the disease.

In 1971, Howard University received $40,000 from the residents of Hartford, Conn. The money, which was used to open the Center for Sickle Cell Anemia at Howard, was raised during a successful campaign conducted by Leonard J. Patricelli, president of Broadcast-Plaza, Inc., WTIC Television Station in that city.

Also, a $100,000 gift was presented to Howard by Howard L. Sanders, president of a New York-based advertising and public relations firm. The $100,000 represented some of the donations received during a four-hour TV telethon conducted in New York by Mr. Sander's organization to raise funds for sickle cell anemia. However, $50,000 of the money given to the University is being held in escrow, pending settlement of a dispute involving the sponsor of the telethon.

The American Oil Company gave $84,052 to Howard for sickle cell research. The money was raised from gasoline sales by 75 cooperating dealers in the Washington Metropolitan area (with a matching contribution by the company) during a 13-week campaign.

The cadets of the United States Military Academy, who once before donated $20,000 to Howard's sickle cell effort, a few months ago gave the University an additional $11,000 raised during concerts sponsored by the cadets.

By the end of 1972, Howard University's sickle cell anemia research and educational programs received a total of $1,497,711.68 from private and Federal sources.

Research Center at Howard

The Center for Sickle Cell Anemia at Howard University was founded in 1971 to coordinate Howard's ongoing sickle cell research and to provide comprehensive screening and genetic counseling (on voluntary basis) services for students at the University and people from the community.

According to Dr. Scott and Dr. Robert L. Hudson, the Deputy Director, the Center is a research-training facility for professional and para-professional personnel whose services are needed to enhance the fight against sickle cell anemia.

The Center has published several pamphlets on the nature of sickle cell anemia, for distribution in the community and for use in public schools. Its diagnostic and consultation resources—part of the University's medical complex—are open to the medical community of the District of Columbia and the surrounding areas.

In June of 1972, the Center's basic services were extended to include a mobile walk-in clinic in the community, to provide free screening and genetic counseling.

As more funds become available, and as the Center acquires adequate working space, its services will increase sharply, Dr. Scott said.

Dr. Marjorie Cates is the Associate Director of the Center. Other Howard doctors associated with the Center and who have helped in its development include:

Dr. William H. Bullock, Dr. V. Bhushan Bhardwaj, Dr. Robert F. Murray Jr., Dr. Angella D. Ferguson, Dr. Ernest L. Hopkins, Dr. Pongrac N. Jilly, Dr. Calvin C. Sampson, Dr. Verle E. Headings, Dr. Allen F. Calvert, Dr. Carlton P. Alexis, Vice President for Health Affairs, and Dr. Marion Mann, Dean of the College of Medicine.

Dr. Scott is perhaps the leading expert of a handful of Black scientists who spent countless hours in laboratories searching for medication that hopefully will bring some relief to victims of sickle cell anemia, a disease which until now the rest of the nation had ignored. (The National Medical Association and the National Association for the Advancement of Colored People are among the few early supporters of the sickle cell disease cause.)

Dr. Scott is regarded as dean of the Black physicians engaged in sickle cell anemia research. His work began more than 20 years ago and has won for him recognition by his colleagues, and a national reputation as one of the country's highly regarded authority on sickle cell disease. He has written more than 50 scientific papers dealing with the clinical delineation of sickle cell anemia syndromes in infants and children; screened more than 20,000 persons for the sickle cell trait; provided genetic counseling for carriers of the trait, and medical care for victims of the malady at the University's medical complex.

"What we are doing here is very significant for Black people in this country, the Caribbean and Africa. It also focuses national and international attention on Howard as a major research center for sickle cell anemia," Dr. Scott said.

"We need financial assistance from alumni, Federal and private agencies, and any legitimate source in order to properly carry out our mission."

He said sickle cell disease indeed is one of the major medical problems facing Blacks, contrary to what other Black Americans are saying.

The National Black Thrust

Elsewhere in the nation, other Black physicians are engaged in research and education like Dr. Scott, but on a smaller scale. Among them is Dr. Allison B. Henderson of Detroit, Mich., who did his first research on sickle cell while serving in the Army in 1945.

Dr. Henderson now conducts most of the research at his office but, when necessary, uses the facilities of Burton Mercy Hospital in Detroit.

Another Black physician in the field of sickle cell research is Dr. Charles F. Whitten, who has done research for more than 10 years.

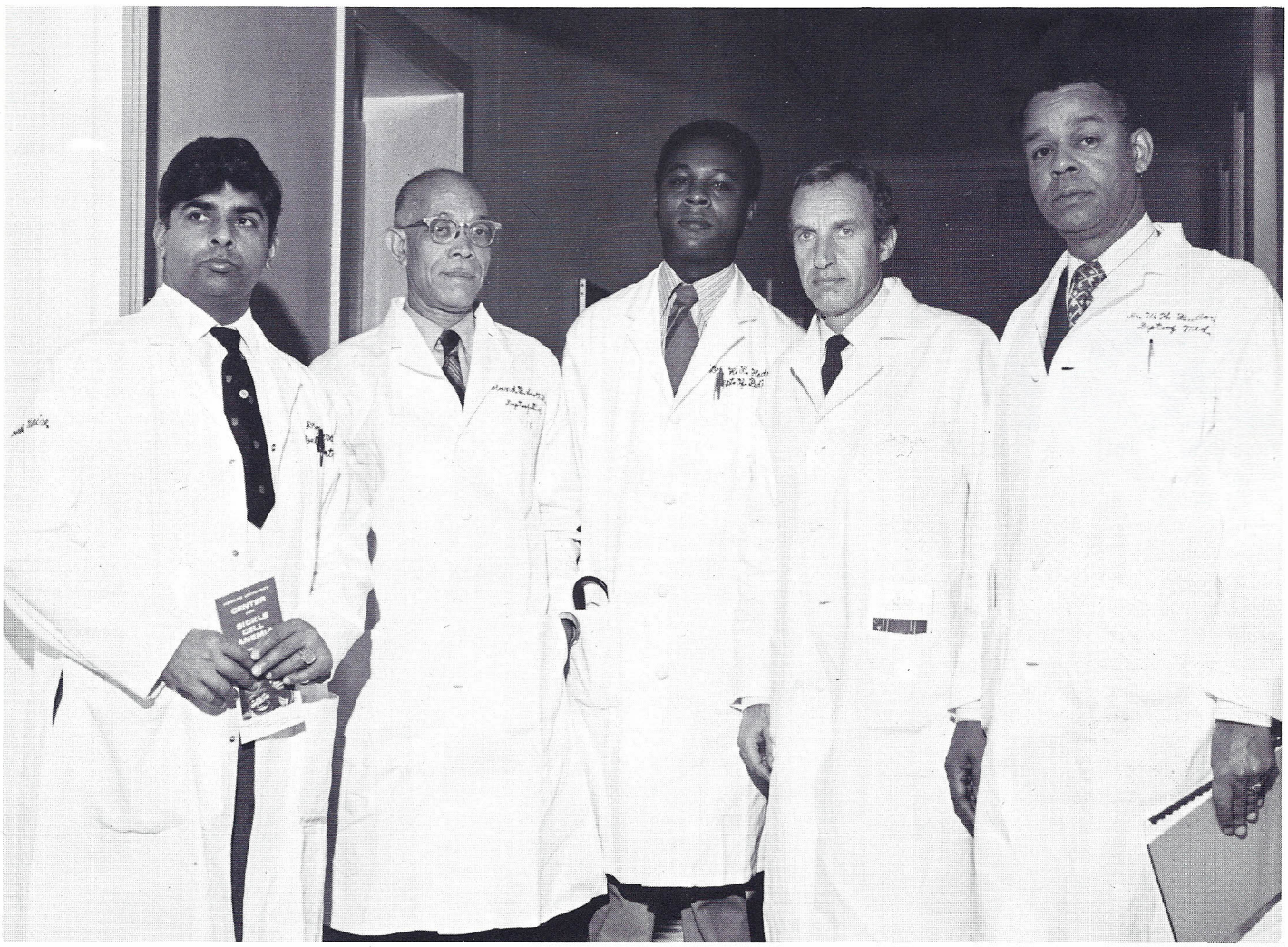
Dr. Whitten also works closely with the Sickle Cell Detection and Information Program in Detroit.

In New York, Dr. Yvette F. Frances does sickle cell research as head of the city's first Sickle Cell Clinic. Also, she is associated with the Foundation for Research and Education in Sickle Cell Disease.

The Foundation, according to Jerry Fowler, a staff member, and a victim of

One of the many functions of the Center for Sickle Cell Anemia is testing new drugs to be used in treating patients. Here, a group of representatives from the Department of Health, Education and Welfare listens as **Dr. William H. Bullock** (holding object) explains urea, a new drug being tested at the Center.

The Team—Left to right, V. Bhushan Bhardwaj, Roland B. Scott, Robert L. Hudson, Pongrac Jilly and William H. Bullock.



sickle cell disease, was formed with donations received from private organizations.

"It is hard to live with the disease without proper medical care," Mr. Fowler said during an interview a few months ago.

Mr. Fowler, 26, received his first treatment when he was nine months old. He has since been hospitalized more than 125 times, received more than 300 blood transfusions, with most of the expense paid by health insurance.

He became associated with the Foundation when it was formed in 1966. Although somewhat similar to the Howard University Center, the Foundation's clinical work is done at five hospitals in New York City, according to Mr. Fowler.

Other Black Voices

The merits of some sickle cell research programs and the sincerity of the persons associated with them especially in the area of fund-raising, are being questioned by Blacks.

There have been charges that some individuals associated with sickle cell anemia programs are interested only in quick financial gain or publicity.

"There are Blacks and whites in the community who are only interested in sickle cell anemia because it is only recently that funds have become available," said William Montgomery of Pittsburgh, Pa., to 1500 delegates attending last year's annual conference of the National Association of Black Social Workers in Nashville, Tenn.

"As funds have become available, numerous people who have never had previous interest in sickle cell have begun to try to develop various kinds of programs. As a result, many of them, we feel, are really in it for their own personal gain rather than in the interest of providing some services to people who have sickle cell."

Elliot Richardson, then Secretary of Health, Education and Welfare, stopped to chat with a young sickle cell anemia patient during a visit to Freedmen's Hospital.

Still other Blacks, for various reasons, view the newly-found popularity of sickle cell as a hoax. They question the motives of some agencies, including the American Red Cross, for example, for instituting sickle cell testing programs.

Professor Naomi Chamberlain, of the University of Rochester, another delegate at the social workers conference, noted how a national organization which previously dealt with hemophiliacs was talking about the possibility of moving into the sickle cell field "because they (the organization) can't find enough hemophiliacs."

Some of the critics, among them physicians and psychiatrists, claim the current popularity of sickle cell is nothing but a scheme by the establishment to demoralize Blacks; another trick to label Black victims of the disease genetically deficient creatures.

Dr. Alyce C. Gullattee, psychiatrist at the College of Medicine at Howard University, has warned Blacks to be aware of the mass screening for sickle cell trait. She is convinced the mass screening of Blacks could be a new means for eugenic control.

Likewise, some Blacks have expressed fear that those with the disease or the trait, some of them potential parents, may be counseled not to have children.

However, individuals associated with sickle cell programs say this won't be allowed to happen.

Responding to this question, Dr. Jackson, coordinator of the Government's sickle cell disease program, said:

"This program is not aimed at counseling anyone not to have children. That's one option they (potential parents) can have. It is up to the couple."

Marjorie A. Costa, director of HEW's National Center for Family Planning Services, said advising victims of sickle cell not to have children would amount to "genocide."

"If I hear of any counselors who are in fact counseling against having children, I think that would, therefore, be genocide."

To forestall such a possibility, Blacks are increasingly calling for more participation in the planning and operation of both Federal and private sickle cell anemia programs.

The lack of adequate participation by Blacks in some programs have in the past come under fire. One such incident involved several Black employees of NIH, who staged a protest and forced the Federal health agency to abandon an in-house sickle cell detection program before it got off the ground. The reason for dissent: Lack of meaningful input into the program by Black employees. (Shortly thereafter, Dr. Jackson was named to head the national program.)

A Quick Look at Past Years

Times have really changed. Sickle cell anemia and its victims are now beginning to get the attention which was lacking only a few years ago.

Writing in the Journal of the American Medical Association (October, 1970), Dr. Robert B. Scott, of the Department of Medicine, Medical College of Virginia, noted:

"In 1967 there were an estimated 1,155 new cases of sickle cell anemia; 1,206 of cystic fibrosis; 813 of muscular dystrophy and 350 of phenylketonuria (congenital metabolic defect in infants). Yet volunteer organizations raised $1.9 million for cystic fibrosis, $7.9 million for muscular dystrophy, but less than $100,000 for sickle cell anemia.

"National Institutes of Health grants for less common hereditary illnesses exceed those for sickle cell anemia," Dr. Scott wrote.

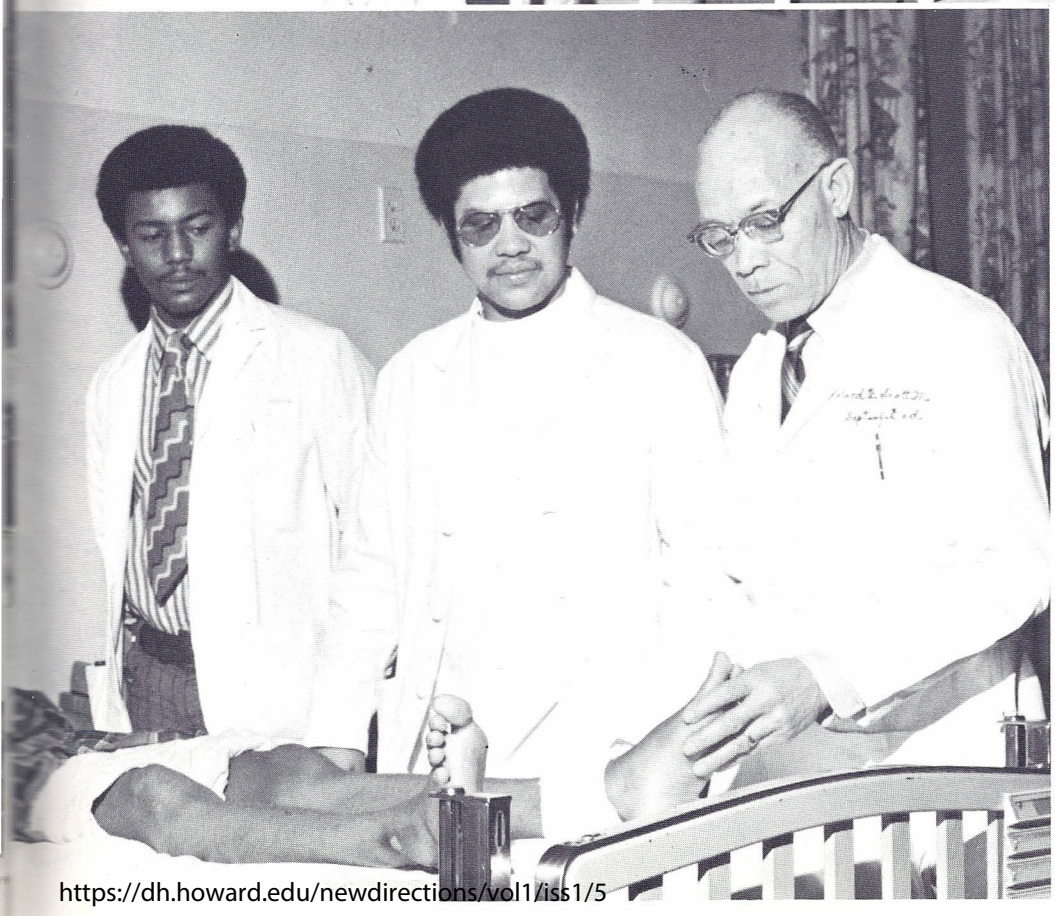
Howard University's Dr. Scott wrote about this neglect in an article published in the January, 1971 issue of the Journal of the National Medical Association, an organization of Black practitioners.

In the article, Dr. Scott made a comparative study between sickle cell disease and other illnesses affecting the predominantly white population in this country. He concluded:

"Vast sums of money have been spent not only from research supported by the Federal Government but also from popular contributions which support national organizations interested in diseases such as muscular dystrophy, special birth defects, hemophilia, etc. On the other hand, the more commonly occurring sickle cell disease afflicting mainly the American Black population has suffered from comparative neglect by the Federal Government and the public at large."

Now, more funds to fight sickle cell anemia have been promised. Whether the various research efforts will produce an effective therapy that will ultimately end the agony and suffering for victims of this malady is another question.

—Abdulkadir N. Said—



Dr. Roland B. Scott (extreme right) often takes time off from his busy schedule to examine patients. Frequently, as in the picture, he takes along medical students.