SICKLE CELL ANEMIA: SOCIAL AND POLITICAL ISSUES

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CHAPTER I

INTRODUCTION

Sickle cell anemia, a hereditary blood disease is cuased by abnormal hemoglobin (oxygen-carrying pigments) in the red blood cells. The disease is characterized by recurrent episodes of pain, paleness of skin, loss of appetite, yellowish color of eyes, symmetrical swelling of both hands and/or feet, and premature death, often in childhood.[1] Of all the genetic diseases to which man is known to be liable, there is probably no other that presents a collection of problems and challenges quite comparable to sickle cell anemia. This disease primarily afflicts blacks, but may also be found in persons of other races.

Sickle cell anemia is one of the best understood diseases as far as its molecular basis is concerned, however, many other facets, including its natural history, are still known to a small degree. Most of the information available is quite general and there are only estimates of the number of patients who have the disease.

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[1]Roland Scott and Althea Kessler, Sickle Cell Anemia and Your Child (Washington, D. C., 1971), p. 8.

While other chronic diseases such as cystic fibrosis, muscular dystrophy, and polio have received widespread attention and a great deal of public and private support, there has been, until recently a small amount of public money made available for research in sickle cell anemia.

In 1971, the United States government under pressure from black organizations and interested medical professionals, began to allocate funds for extensive research of the disease. In 1972, Congress approved a $115 million--3 year program for sickle cell anemia research, and several cities and states have instituted screening programs, or passed legislation requiring it. In addition to these public moves, a number of private foundations have been formed to raise money, promote research and provide information on the disease.[1]

History and Background

In the case of sickle cell anemia, the discussion seems to require at least a brief review of the history of the disease state itself in the United States. This, of course, must reflect the context of the now much publicized "crisis" in medical care.

Sickle cell anemia is a hereditary blood disease that affects more than 2 million persons widely spread throughout the world. It is generally accepted that sickle cell disease

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[1]Congressional Record 117 (February 18, 1971), pp. 3019-21.

originated in Africa thousands of years ago. The gene for sickle cell anemia is believed to have been introduced to other parts of the world by migration of early Africans. The disease is found not only in Africa but also in the West Indies, Central and South America, the Middle East, in the Mediterranean region, India, and in American states with sizeable black populations. The beginning of the sickle cell disease is today still a matter of discussion. As with other genetic diseases it came about because of a gene mutation, or change, during man's evolution.[1]

The first description of the distinquishing characteristics of sickle cell anemia was given in the medical literature in 1910 by a Chicago clinician, Dr. James B. Herrick. He wrote about a twenty year old West Indian student whose symptoms involved "a secondary anemia... strikingly atypical in the large number of nucleated red cells of the normoblastic (immature) type and in the tendency of the erythrocytes (red blood cells) to assume a slender, sickle-like shape."[2] This peripheral blood smears of the patient showed that in addition to the normal, round red blood cells, a number of cells had an unusual sickle shape

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[1]Anthony Cerami and Elsie Washington, Sickle Cell Anemia (New York, 1974), p. 90.

[2]Michael Michaelson, "Sickle Cell Anemia: An Interesting Pathology," Ramparts Magazine 10 (October 1971): 53.

and also fewer cells.[1]

Herrick waited six years before he published his original observations because he was concerned that the patient did not actually have a new disease. Previously, the pain of sickle cell had been thought to be arthritis, rheumatic heart disease or the result of syphilis.[2]

Other clinicians reported on patients with similar symptoms and substantiated Herrick's original observations. Dr. Emmel in 1917 was the first to use the term "sickle cells" repeatedly, Mason in 1922, the first to refer to "sickle cell anemia" and Graham and McCarthy in 1926, the first to point out that this anomaly of red cells afflicted only blacks.[3]

E. Hahn and E. Gillespie made the next major breakthrough in the understanding of the sickle cell disease in 1927. They found that when oxygen was removed from red blood cells the cells sickled, that is, became irregular in shape. However, when oxygen was transferred back to the sickled cells they would regain their original, doughnut-like shape.[4]

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[1]Cerami, Sickle Cell Anemia, p. 3.

[2]Ibid., p. 3.

[3]Michaelson, "Interesting Pathology," p. 54.

[4]Cerami, Sickle Cell Anemia, p. 3.

It was therefore theorized that:

> When cells give up their oxygen and sickled in the capillaries, the abnormal, jagged shape inhibits their continuing passage through the capillaries. The entrapped sickle cells prevent the passage of other red cells which in turn sickle and further clog the capillaries. If the blockage is not alleviated, the surrounding tissues will die because they do not receive its functional and vital supply of oxygen.[1]

Following the discoveries of Hahn and Gillespie, Linus Pauling in 1949 described the molecular basis for this disease. Through electrophoresis he showed the existence of a difference in the red cells from a patient with sickle cell disease compared to the red cells of a normal patient. This hemoglobin was identified as Hemoglobin S, in contrast to the normal adult hemoglobin called Hemoglobin A. In addition, Pauling showed that the carriers of the genetic defect, trait individuals, had both Hemoglobin A and Hemoglobin S in their cells. Since Pauling's observations, more than 150 abnormal hemoglobins, or hemoglobinopathies, have been described by other scientists.[2]

At the time when Pauling worked on the chemistry of the defect, a human geneticist, James Van Neel, showed that:

> The sickling gene behaves lide the ABO blood group genes, as co-dominant with the normal gene. That is, individuals with one normal and one sickling gene are generally healthy but have the sickling trait, which can be tested through their blood and detected. These are what we now call carriers or, as geneticists say, heterozygotes. If two carriers

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[1] Ibid., p. 3.

[2] Ibid., p. 4.

> marry, they may have children who have two sickling genes; those children have sickle cell anemia. Each child with sickle cell anemia must have had parents who are both carriers of the strain.[1]

In the early 1950's, Vernon Ingram, a biochemist, devised a method of pinpointing the precise nature of the molecular abnormality. Through this method he determined that all of the amino acids (building blocks of proteins) of the Hemoglobin S and Hemoglobin A molecules were the same except for one. The minute difference in the complicated hemoglobin molecule leads to the sickling and the pathological state that affects every organ of the patient's body.[2]

The genetic and molecular character of sickle cell anemia has made it important in the evolution of medicine in the last 50 years.

Statement of the Problem

Sickle cell anemia as a national health care problem has been neglected for approximately 60 years. For so long it has been considered an "interesting pathology" to doctors and other medical professionals. Suddenly, however, sickle cell anemia has received much publicity and funds from both public and private sectors of the society. This particular health problem has been discussed

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[1]Jessyca R. Gaver, Sickle Cell Disease--Its Tragedy and Its Treatment (New York, 1972), p. 15.

[2]Cerami, Sickle Cell Anemia, p. 4.

and debated at various black professional meetings, such as the National Medical Association, the National Association of Black Social Workers, and black political organizations and the like. As a result of this sudden interest the black community has become increasingly suspicious. In many ways, it is conceivable that this health issue has many implications which are just becoming apparent. Most blacks believe that these crash programs for the disease is not medical in the strickest sense, yet seem to suggest both social and political undertones. Therefore, this study will be primarily concerned with identifying and critically analyzing the major socio-political issues associated with the sickle cell anemia controversy.

Review of the Literature

Sickle cell anemia as is well known is largely confined to black Africans and derivative populations, such as the Negro of the New World. Livingstone has summarized the frequency of sickling in these populations based on extensive surveys. In Africa itself, the gene has an uneven distribution, which varies from a heterozygote frequency to near zero in some tribes to a frequency of 40 percent in others.[1] Migration and selection of complex interaction

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[1]Frank B. Livingstone, Abnormal Hemoglobins in Human Population.(Chicago, 1967), pp. ix and 470.

has produced the observed pattern of distribution.

Because of the loss of tribal identity enforced by slavery, the gene frequency is more uniform throughout the Negro population. In the Americas, in part because of the fact that the average Negro is some 20 percent Caucasian in his ancestry, in part because of altered selective pressures, the frequency of the sickle cell trait appears to be lower than in the tribes or regions of origin of the American Negro. The figures for the Negroes of Latin America are extremely variable, to a first approximation the estimate is about 8 percent. For the United States Negro, the frequency of classical sickle cell disease among newborn infants prior to the action of selective mortality should be approximately 2.7 per 1,000; and for the Negro of Latin America, the expectation is 1.7 per 1,000 births. Among a population of somewhat more than 20,000,000 Negroes in the United States, the expected number of cases is 54,000; but because of the differential mortality, the actual number is perhaps one half to two thirds of this.[1]

Song in a monograph for reference work, gives descriptions and discussions on this disease based on a total of 422 cases of sickle cell disease, 111 patients with homozygous sickle cell disease (sickle cell anemia), 306 cases of heterozygous sickle cell disease (sickle cell trait); and five patients with sickle cell homoglobin C disease.

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[1]Ibid., p. 471.

The information provided in this monography is extremely technical and detailed covering both medical and clinical facets of the disease.[1]

In the same vein Abramson, Bertles, and Wethers edits a book based on a symposium designed to consider in depth the important disease complex. The book presents a reexamination and reassessment of the genetic abnormality. It further discusses the organization of specialized clinics, management, comprehensive health care programs and the like.[2]

Scott in several studies have illustrated the relative incidence of sickle cell disease compared with that of other similarly serious major childhood illnesses. In addition, these data on the incidence of sickle cell anemia show that the disease occur in about one in 500 black births in the United States. Some other exclusive conditions occur almost extentively among Caucasian children. Examples are, cystic fibrosis and phenylketonuria. In years past the publicity and priority given these diseases have not necessarily reflected their relative incidence.[3]

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[1]Joseph Song, Pathology of Sickle Cell Disease (Illinois, 1971), pp. 1-548.

[2]Harold Abramson, John F. Bertles, and Doris L. Wethers, Sickle Cell Disease (St. Louis, 1973), pp. 1-350.

[3]Robert B. Scott, "Health Care Priority and Sickle Cell Anemia," Journal of the American Medical Association 214 (October 26, 1970): 731 and Robert B. Scott, "Sickle cell Anemia--High Prevalence and Low Priority," New England Journal of Medicine 282 (1970): 164.

The 1970 census data shows the prevalence among both Negro and Caucasian children of sickle cell anemia, childhood diabetes, childhood nephrosis, diabetes mellitus and cystic fibrosis. In Philadelphia, sickle cell anemia is at least as prevalent as diabetes mellitus and exceeds the prevalence of cystic fiborsis severalfold. In cities with increasing proportion of Negro populations, sickle cell anemia becomes inordinately more prevalent than these other chronic illnesses. Outside the South, virtually 95 percent of all Negroes live in metropolitan areas.[1]

Based on the high incidence of the disease among black children it is presumed that the black community would be aware of this condition. This was ascertained in Richmond, Virginia, in the beginning of the Virginia Sickle Cell Anemia Awareness Program (VaSCAP) by conducting an inclusive survey of adult Negroes. There were 1,457 individuals who were asked whether they had ever heard of sickle cell anemia. If an affirmative answer was given, they were then asked in 1968 only three out of ten adult Negroes had ever heard of sickle cell anemia, despite its high prevalence and obvious importance in the community. In the survey it was also recognized that those individuals who had heard of sickle cell anemia still had little or no understanding of the nature of the

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[1]U. S. Department of Commerce, Bureau of Census, The Social and Economic Status of Negroes in the United States--Current Population Reports, P-23, no. 38 (Washington, D. C., 1971), p. 117.

condition.[1]

Two medical students, Greenberg and Wethers, conducted a study which involved the screening of 650 head-start children in Boston for the sickle cell trait. This study corraborates the Lane and Scott survey. It was found that over half of these children had the sickle cell trait and less than half of the parents had never heard of this disease. Those parents who had heard of the disease, did not know that the disease was hereditary.[2]

The degree of neglect with regard to this national health care problem has been well documented. However, it now seems that the disregard for this problem is over. For the first time funds for research in significant amounts are being made available by the federal government.[3]

Through the increase in funds, counseling and mass screening programs have been developed in the black community. Basic authorization to conduct screening is, how-

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[1]John C. Lane and Robert B. Scott, "Awareness of Sickle Cell Anemia Among Negroes in Richmond, Virginia," Public Health Reports 84 (November 1969): 949-957.

[2]Mortimer S. Greenberg and Doris L. Wethers, "Need to Identify Sickle Trait," The New England Journal of Medicine 282 (March 21, 1970): 629-636.

[3]"Facts About Sickle Cell Anemia," National Foundation of the March of Dimes (White Plains, 1970) and William W. Zueler, "Pediatric Perceptions: The Pediatrician and the Species: Some Implications of our Acheivements," Pediatrics 47 (1971): 339.

ever, doubtless one of the most critical issues today, mainly because all such programs are subject to enabling legislation or administrative ruling based on general or specific statutes. Within the years 1971-1972, several states enacted laws to permit screening for sickle cell disease referrals and treatment as well as counseling.[1]

Parker states that the ethical and legal aspects of genetic counseling are "still a virgin field" calling for study by scientists and lawyers alike. According to Parker, perhaps two trends can be discerned: (1) screening laws, as for phenylketonuria, probably as an overreaction to the belief that the diagnosis and treatment of the condition are scientifically certain, and (2) general eugenic legislation, going back many years, (a) to reduce undirable births, on the one hand, and (b) to permit possible or desired births, as perhaps through artificial insemination, on the other hand.[2]

Data Collection

The data instrumental in the development of this study were made available through secondary sources directly and indirectly related to its purpose. Most of the data

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[1]Indian Acts of 1971, P.L. paragraph 1 (1971), p. 824; Massachusetts Chapter 491, Special Act 105 (July 1, 1971); and Connecticut House of Representatives, Bill 1519, (1971).

[2]Carlton Parker, "Some Ethical and Legal Aspects of Genetic Counseling, Birth Defects (1970), p. 52.

collected was derived primarily from studies published in leading medical journals, magazines and various health reports. Other materials also implemented in the construction of this study consist of books relating to sickle cell anemia. In addition, several legal and governmental documents and census data were employed. Data related directly to this subject matter appeared to be rather sparse. This is possibly due to the fact that just within the last three or four years that funds have been received for research in sickle cell disease.

Methodology

The method involved in this study is basically of an exploratory nature which is centered around a review of related pertinent literature. The data collected for this study further involved identification and critical analysis of the variables under investigation.

CHAPTER II

CLINICAL MANIFESTATIONS[1]

Sickle cell anemia has been referred to as a "killer" and "crippling" disease over past years. Some children have a stormy course with many crises and complications that result in frequent hospitalizations and an early death. Others have a more benign form of the disease that allows them to lead normal lives. Thus, in order to become fully comprehensive of the severity of sickle cell disease, it is imperative to have a clear conception of the manifestations of this disease. There are symptoms presented in common by most persons who have sickle cell disease.

The most universal of these symptoms is an *anemia* which is a deficiency in the number of red blood cells. The degree of the anemia varies from patient to patient and ranges from moderate to severe.

There are three common clinical ways to measure the degree of an anemia. These are: by determining the hematocrit, by measuring the concentration of hemoglobin or by

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[1]The source for the technical discussions on sickle cell anemia are found in Anthony Cerami and Elsie Washington, *Sickle Cell Anemia* (pp. 8-17). All italics in this chapter were made by the writer of this study.

counting the actual number of red blood cells. A small blood sample of the patient is centrifuged and the volume taken up by the red bloods compared to the total volume of the blood is the hemacrit.

Anemias can be caused by an underproduction of red blood cells, early destruction of these cells, or a combination of the two. The anemia of sickle cell disease is a result of the rapid destruction of the sickled red blood cells. The life span of a red cell containing sickle hemoglobin (Hemoglobin S) is 30 days, less than a third of the 120 day life span of normal red cells.

In a patient with sickle cell anemia, the bone marrow, which is the location of the production of red cells, expands drastically to keep up with this rapid destruction of the cells. In a healthy, mature adult, red cell production is primarily confined only to the axial skeleton, that is, the bones in the trunk of the body. The other bones of the body, such as those in the arms and the legs, are filled with fat cells rather than marrow. It has been estimated that a normal adult produces 900 billion red cells a day. Patients with sickle cell disease, as a result, of the anemia, produce seven to ten times this number of cells. Consequently, every hollow bone in the body is utilized for red cell production. In spite of this desperate effort, cell production cannot equal cell destruction.

A rare, but serious complication in patients with sickle cell disease is a hematological crisis which occurs

when the bone marrow ceases to produce cells. It is not known exactly why this occurs but it is thought that these crises are precipitated by viral infections. Such cessations of the production of red blood cells in a normal person does not lead to any serious consequences. However, because of the continual and rapid destruction of the red cells without replacement in a patient with sickle cell disease, there is a precipitous drop in the hematocrit which can evolve into a life-threatening situation.

Because of the reduction in the number of red cells, the muscles and other tissues of the patient receive a marginal supply of oxygen. If he exerts himself physically, he tires quickly because his muscles develop what is termed an oxygen debt. At this time, the muscles will produce large amounts of lactic acid, a waste product that is responsible for the pain of muscle fatigue. Another manifestation of the anemia is the occurence of pale lips, gums and fingernail beds.

When the red blood cell is destroyed by special "scavenger" cells, hemoglobin is separated into heme and globin components. The globin is further cleaved into individual amino acids which can then be used for the assemblage for other proteins. The heme ring is split into iron, carbon monoxide and bilirubin. The iron is conserved and used for the production of new hemoglobin. The production of carbon monoxide, a toxic gas, by this reaction is unique, representing the only reaction of its turnover by the amount of carbon

monoxide evolved in the expired air. This expired amount of carbon monoxide even in a patient with sickle cell disease is comparatively small and is similar to the levels of a smoker is exposed to.

The bilirubin that results from the breakdown of the heme molecules is a red pigment. It is carried in the plasma (the fluid, non-cellular portion of the blood) where it is excreted into liver bile. The bile then goes into the gall bladder where it is concentrated. Because bilirubin is not very soluble it forms stones that can obstruct the bile ducts and lead to gall bladder disease. In patients with sickle cell disease the liver is unable to accomodate the increased production of bilirubin and there is, as a consequence, an increased level of plasma bilirubin. As a patient ages, the liver functions less efficiently, and the plasma bilirubin rises to even higher levels. This increase causes jaundice, a yellow staining of the skin and the whites of the eyes.

After a period of time, as a result of the anemia and the sickling process, the heart undergoes a number of changes. The most frequent modification is an enlargement of the heart. This can be quite dramatic, with some patients having a heart two to three times the normal size. A large number of the patients also develop heart murmurs that in the past have been misinterpreted as being caused by congenital or rheumatic heart disease. The sounds of a heart murmur are the result of blood spurting back into the heart chambers, against the normal flow, because of improper closing of the

heart valves. In addition, patients with sickle cell disease frequently develop arrhythmias, an irregularity of the heart beats.

As a result of these pathological changes in the heart, older patients with sickle cell disease frequently suffer from heart insufficiency. Subsequent complications are swollen extremeties (edema), enlarged, tender liver engorged veins and pulmonary congestion.

Another common effect of the sickling process is chronic leg ulcers. Seen mainly in young adults, these ulcers occur in the ankle region and are slow to heal because blood circulation is poor in this part of the body. Agitation from walking and accidental jolts that re-open the wounds further hinder healing. Skin grafts to these ulcers have been tried but, for the most part, have not been successful. They are best managed by immobilizing the ankle and keeping the wounds clean.

The course of sickle cell disease also adversely affects the urogenital system. One of the most commonly observed aspects of this is a delayed sexual maturation of the external genitalia and secondary sex characteristics, i.e., beard growth, breast development.

In women with sickle cell disease, pregnancy can be life-threatening, for all the symptoms and complications of the disease are intensified by the extra burden of nourishing the fetus. It has become common practice to transfuse pregnant patients with normal blood throughout the pregnancy

and hospitalize them in the final stages.

A rare complication that male patients suffer is called a priapism. Large numbers of sickled cells become entrapped in the penis and result in a painful erection that can last for hours, or days. Operations to remove the sickled cells have provided temporary amelioration, but in most cases the priaspism returns. Eventually white blood cells remove the entrapped cells and surrounding dead erectile tissue and the priapism regresses. However, there is a high incidence of impotence following such episodes. Priapisms have also been observed in rare instances in sickle trait individuals.

Another occasional symptom that sickle traits individuals have in common with sickle cell disease patients is hematuria, or blood in the urine. The reason for this bleeding is not understood, but might reflect damage to the kidney because of the sickling process. The sickling of red cells also leads to a decreased ability of the kidneys to concentrate urine, one of its prime functions. This function is extremely important during peroids of restricted water intake because the kidneys will then conserve water and produce a more concentrated urine in a smaller volume. A diminished water intake in a patient with sickle cell disease can be dangerous since he cannot concentrate his urine and conserve water. His normal volume of urine secretion continues and he becomes dehydrated, a condition believed to precipitate the crisis.

Although sickling can occur in the vascular of the brain, leading to serious neurological symptoms, the incidence of such complications is infrequent. When cerebrovascular occlusion does occur patients exhibit neurological manifestations similar to those observed with patients suffering from strokes. These may include drowsiness, paralysis, transitory or permanent blindness, aphasia and generalized convulsions.

The skeletal system is frequently affected in the course of sickle cell disease. This is evidenced by: (1) an increase in the size of marrow cavities, (2) the death of the bones because of a blockage of the blood supply, and (3) an increased incidence of osteomyelitis, which is bacterial infection of the bone.

The bacteria most commonly causing osteomyelitis in patients with sickle cell disease is salmonella. It is not known exactly why these patients are so susceptible to this organism. Some investigators have proposed that there is a defect in the ability of the patient's white blood cells to attack and destroy the invading micro-organisms. This destruction of the bone can permanently damage the system, particularly in the head of the femur (thigh) and humerus (upper arm) bones, and in many cases the patient is crippled.

A common bone abnormality found in children during the first two years of life is a swelling of the extremeties called the hand-foot syndrome. This swelling is a result of tissue death in the bones.

Another manifestation of sickling is arthralgia, or pain in the joints. This arthralgia is common and constitutes one of the major complaints of patients with the disease.

The disease manifestations described create a chronic destruction of the patient's organs. In addition, there are peroids of excruciating pain called crises. This pain is usually experienced in the abdomen, chest and joints, and it is believed to be caused by the entrapment of sickled cells and subsequent clogging of the capillaries of the tissues involved. In some patients a crisis lasts for several hours, in others, it continues for days. It is not known specifically what precipitates all crises, but exposure to cold, to heat, viral or bacterial infection, and loss of body water have been implicated as causes. The severity of the pain varies from mild, requiring only aspirin, to severe, which requires hospitalization. During hospitalization for a severe crisis, patients are administered intravenous fluids to combat possible dehydration caused by a diminished fluid intake. They are also given narcotics to alleviate the pain. The incidence of crises in a patient is unpredictable. He may suffer many of them at one stage in his life, and relatively few in another. Most patients average two to three crises per year.

Usually, these episodes of pain are resolved within a week. However, a crisis can lead to permanent damage to the organs, or occasionally, to the death of the patient. A typical crisis leaves the patient in a state of weakness and exhaustion that requires two to three weeks for full recovery.

CHAPTER III

SOCIAL ASPECTS OF SICKLE CELL ANEMIA

The Question of Priorities for the Community

Sickle cell anemia is considered to be one of the most common long-term childhood diseases of black children. It is estimated that half of the children who are afflicted die before the age of twenty. The disease occurs in about one in 500 black births according to data on the incidence of the disease. The incidence of sickle cell anemia is higher than other childhood diseases such as phenylketonuria, diabetes mellitus, cystic fibrosis, and acute leukemia.[1] It is worth noting that the victims of these diseases are primarily white which is illustrated in Table 1. Furthermore according to the one to 500 births, it is estimated that there are at least 50,000 black persons in the United States with sickle cell anemia. Based on the data made available in Table 1 it would be appropriate to assume that sickle cell anemia should be one of the most important health concerns of our day.[2]

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[1]Scott, "Health Care Priority," p. 731.

[2]Ibid., p. 732.

TABLE 1

ESTIMATED NEW CASES IN U. S., 1967

	Sickle Cell Anemia	Cystic Fibrosis	Muscular Dystrophy	Phenyl-ketonuria
Negro	1,142	17	114	0
White	13	1,189	699	350
Total	1,155	1,206	813	350

SOURCE: Robert B. Scott, "Health Care Priority and Sickle Cell Anemia," Journal of the American Medical Association 214 (October 26, 1970): 732.

Table 2 illustrates estimates of chosen long-term illnesses in four cities with a large black population. Sickle cell anemia is apparently more prevalent than the other childhood diseases in all four cities. The 1970 census data indicated that there are thirty cities with a black population of 88,000 or more.[1] It is estimated that this number will increase to fifty by 1980. These data tend to suggest that sickle cell anemia will continue to be a major community health problem in urban areas.

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[1]U. S. Department of Commerce, Bureau of Census, The Social and Economic Status of Negroes in the United States--Current Population Reports, P-23, no. 38 (Washington, D. C., 1971), p. 17.

TABLE 2

PREVALENCE ESTIMATES: SELECTED LONG-TERM ILLNESSES (WHITE AND BLACK CASES/100,000 CHILDREN) IN FOUR CITIES

Diagnosis	Philadelphia 32% Black	Richmond, Va. 44% Black	Detroit 47% Black	Washington, D. C. 68% Black
Sickle cell anemia	61	84	89	129
Diabetes mellitus	50	47	46	39
Nephrosis	15	16	16	16
Cystic fibrosis	10	8	8	5

SOURCE: Robert B. Scott, "Health Care Priority and Sickle Cell Anemia," Journal of the American Medical Association 214 (October 26, 1970): 733.

With regard to the high incidence of the disease among black children, one would expect the black community to be aware of this condition. In order to determine the level of awareness of sickle cell anemia in the black community, a survey was conducted of 1,457 adults in Richmond, Virginia. It was concluded from the survey that:

> Only 30 percent of those questioned had heard of this disease. Of those who had heard of it, many apparently did not understand the nature of the illness. Awareness of sickle cell anemia was closely related to the educational level of the persons surveyed.[1]

The conclusions derived from this study has received widespread attention from Congressmen, health officials, and grassroot organizations. Another study involved the screening of headstart children in Boston for the sickle cell trait.[2] It was found that 7.3 percent of 650 tested had the trait and that 47 percent of the parents of these children had heard of the disease. The parents of these children were not aware that the trait was transmitted from parents to the child. These data, therefore corraborate the previous study of the level of awareness. Public knowledge of sickle cell anemia is not widely spread, eventhough it is a major childhood illness among black children.

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[1]Lane and Scott, "Awareness of Sickle Cell Anemia," p. 952.

[2]Greenberg and Wethers, "Need to Identify Sickle Trait," p. 629-30.

In view of the facts, even the editors of the conservative Journal of the American Medical Association were forced to conclude, "clearly focuses a spotlight on public failure to recognize the importance of combating sickle cell anemia."[1]

Furthermore, the journal found it "incredible... that very few people in the black population at large have been offered pertinent information about sickle cell anemia and the mode of its transmission."[2] On the other hand the Black Panthers find it to be expected and have launched a major drive through their People's Free Clinics to provide the black community with information about the disease along with free diagnostic blood tests.

This disease has a significant incidence in the black community which the public health data illustrates. In addition,

> although the disease is found in one of five hundred black babies, and although these children survive only an average of twenty years, and although very much more is understood of the genetics, chemistry and epidemiology of sickle cell than of other serious diseases of childhood, physicians and researchers within the white establishment have virtually ignored it.[3]

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[1]"A Commentary on Sickle Cell Anemia," Journal of the American Medical Association 215 (1970): 942.

[2]Ibid., p. 942.

[3]Michaelson, "Interesting Pathology," p. 53.

Furthermore, the private and public sectors of our society have not provided much financial support for research and development for this condition. The data in Table 1 indicates that in 1967 there were an estimated 1,155 new cases of sickle cell anemia, 1,206 of cystic fibrosis, and 813 of muscular dystrophy. Yet, "volunteer organizations raised raised $1.9 million for cystic fibrosis, $7.9 million for muscular dystrophy, but less than $100,000 for sickle cell anemia."[1] Although these figures have changed especially in the past few years, the fact remains that sickle cell anemia has not received the same level of private financial support for research and education as other childhood diseases. Until recently, the federal government, through the research contract and grant mechanisms of the National Institutes of Health (NIH) has not provided the level of assistance for sickle cell anemia as would be suggested by the data on the incidence of the disease. For instance, in 1968, NIH gave only 22 grants for sickle cell anemia compared to 92 for acute leukemia, 41 for phenylketonuria or 23 for glycogenoses, an exceedingly rare condition.[2] In the 1972 fiscal year, there were 72 contracts and grants for sickle cell anemia sponsored by NIH.[3] In the imme-

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[1]Scott, "Health Care Priority," p. 73.

[2]Ibid., p. 733.

[3]Rudolph E. Jackson, "Goals of Federal Programs in Sickle Cell Anemia," Meadowbrook Staff Journal (Spring 1973): 24.

diate year prior to 1970 it was estimated that only $1 million per year was spent by the federal government for sickle cell anemia related problems. By 1972, this sum had increased to $10 million.[1] Through the passage of the Sickle Cell Anemia Control Act the financial picture for sickle cell anemia research and treatment changed drastically. Under the bill, the federal government established its own nationwide program under NIH which authorizes the expenditure of $25 million for 1973, $40 million for 1974, and $50 million for 1975.[2]

Many blacks are beginning to question the importance and attention given to the disease, because of the financial support provided by the federal government as well as private foundations for sickle cell anemia research and education. The following statement was issued by the Center for Black Education:

> How important is this disease to black people? The answer is that while sickle cell anemia is a crippler and a killer well-deserving of a cure, there are many things which more commonly afflict black people in America. There is lead poisoning which millions of black children living in run-down slum homes contract from paint peeling off the walls. There is malnutrition from inadequate diets of many black people in the rural southern part of the country as well as in the cities. There is dope addiction and hypertension which is common in the crowded cities of Chicago, New York and Los Angeles."[3]

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[1]Ibid., p. 17.

[2]Public Law 92-294 (92nd Congress, S.2676, May 16, 1972), p. 2.

[3]Muhammad Speaks, January 21, 1971, p. 4.

While these assertions could be labeled rhetoric, governmental statistics tend to support them. For instance, maternal mortality is three times higher for blacks than for whites; and infant mortality is also greater for blacks.[1] It is generally known that the hypertension death rate is higher for blacks than whites. In 1967 it was estimated that this disease killed 13,500 blacks compared to 340 for sickle cell anemia.[2] It has been estimated that 80-85 percent of lead poisoning victims in the United States are nonwhite.[3] The data from a nutritional survey of preschool children from impoverished black families in Memphis showed that anemia was common.[4] It was also found that on the anthropometric charts that half of the children were below the 25th percentile for height and weight. The researchers concluded that lack of food was the main cause of growth retardation and anemia.

It was debated that the differential mortality rates for specific diseases are highest for the infectious disease

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[1]U. S. Department of Commerce, Bureau of Census, p. 98.

[2]U. S. Public Health Service, Vital Statistics of United States, Vol II, Mortality, Part B (Washington, D. C., 1969), pp. 1-63.

[3]Fact Sheet on Institutional Racism (New York, 1971), p. 3.

[4]Paul Zee et. al., "Nutrition and Poverty in Preschool Children," Journal of the American Medical Association 213 (August 3, 1970): 739.

group. This observation leads to the conclusion that:

> By and large, medical science has not made a dramatic reduction in mortality due to malignant disease even in the white community so the lack of medical care does not affect the course of these diseases markedly from one racial group to the next. Medical science, however, found cures and controls for most infectious diseases, which have greatly reduced their mortality rates in the white community. But the health institutions have failed to effectively extend this significant medical progress to the black community.[1]

Myths and Misconceptions

In the last three years, sickle cell anemia, a previously little-known disease has captured the attention of the United States President, television scriptwriters, fund raisers, legislators, medical and social workers and, especially, the country's black poulation. As with other "popular" diseases, there is a great deal of misunderstanding and misapprehension about its nature which is due primarily to inadequate public information about the disease.

Sickle cell anemia has been headlined as a "black scourge" and a "killer disease." Because sickle cell anemia is hereditary, these terms are considered by some to imply physical inferiority or that "bad blood" is inherent in blacks. Without education and counseling about the disease, this implication can be harmful. Many blacks have found that they have the trait and have felt frightened and stigmatized.

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[1]Louis L. Knowles and Kenneth Prewitt, eds., Institutional Racism in America (New York, 1969), p. 98.

They also panic when they discover that they have the trait because of ths killer image attached to the disease. It is felt that this image exaggerates the character of sickle cell anemia. While the disease is debilitating and, more often than not, shortens apparently patient's life expectancy, there are mild forms of the disease. In addition, scientists have identified several substances, such as urea and cynate, which have shown some successes in helping patients to cope with the sickle cell crisis.[1] However, it shoul be noted that when scientists announced that urea showed some promising results, two newspapers and a magazine came out with headlines saying, "cure found for sickle cell." Of course, this type of information reporting leads to confusion and unrealistic expectations that a cure is on the horizon.

Some medical literature has also promoted myths and misconceptions about the conditions associated with sickle cell anemia. For instance, some brochures issued by the National Institutes of Health has equated sickle cell trait with sickle cell anemia.[2] In several situations, medical literature states that few sickle cell anemia patients live beyond their 40's, but these assertions are based on rather limited studies; for many patients live well beyond that age.

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[1]Gaver, Its Tragedy and Its Treatment, pp. 75-83.

[2]James E. Bowman, "Mediolegal Social and Ethical Aspects of Sickle Cell Programs," Meadowbrook Staff Journal (Spring 1973): 34.

What is significant is that the seriousness of the disease can be influenced by other factors such as malnutrition and generally poor living standards. Of course, these factors are clearly related to the social structure of our society as they impact on the delivery of health services to the black community.[1]

Furthermore, some people equate the sickle cell trait with the disease and this has caused undue harm and magnification of the sickle cell problem far beyond reality. The confusion between the trait and the disease can be observed in the language of the National Sickle Cell Anemia Control Act. For example, the first list of the law reads, "... sickle cell anemia is a debilitating, inheritable disease that affects approximately two million American citizens and has been largely neglected."[2] This grievous error is most unfortunate in that the law sets national policy for this important problem. Some state laws have also tended to confuse the trait and the disease. One consequence of this confusion is that there have been charges that jobs and insurance have been denied to persons who carry the

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[1]Ibid., pp. 33, 34, and 39 and Stanley Smith, "The Socio-psychological Aspects of Sickle Cell Anemia," Meadowbrook Staff Journal (Spring 1973): 4-10.

[2]Public Law 92-294 (92nd Congress, S2676, May 16, 1972), p. 1.

trait.[1] Apparently, there is little or no evidence that trait carriers have a higher risk of disease or shorter than normal lifespan or have not been productive in their jobs. It has been concluded that "careful, controlled studies of carriers of the trait are few and far between, so the matter is clouded by a slew of impressions and erroneous notions."[2]

Another myth promoted by public misinformation is the fallacy that sickle cell disease is a black man's disorder. Relying on literature published by leading private and public agencies, the United Klans of America has been circulating materials which attempt to associate black people with monkeys and apes in arguing that sickle cell anemia is common to only blacks and these animals. As stated, both the trait and the disease are found not only in Africans and their descendants, but also in persons of various other races.[3] Due to racial mixing, it has been estimated that 20 percent of the United States' whites have African genes. Therefore,

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[1]Barbara Culliton, "Sickle Cell Anemia: The Route from Obscurity to Prominence," Science 178 (October 13, 1972): 142 and Thomas M. Powledge, "New Ghetto Hustle," Saturday Review of Sciences 1 (February 1973): 38-40 and 45-49.

[2]Culliton, "Route from Obscurity to Prominence," p. 142.

[3]Jane S. Fin-Fu, Sickle Cell Anemia: A Review of the Literature (Washington, D. C., 1965) and Livingstone, Abnormal Hemoglobins, p. 472.

it follows that some whites have the trait and some also have the disease. Stigmatizing black people as having a genetic disorder must be seen in terms of how the concept "black" has been viewed in Western society. It has been argued that:

> The denotation and connotation of black carry with it a long and persistent history of negativism and pathology in Western civilization.... The association of blacks with sickle cell anemia pervades the minds and thoughts of the average American. Diagnosis treatment and research resources and policies of implementation will be to a large extent contingent on how these associations are conceptualized.[1]

Publicity about the racial and genetic factors of sickle cell anemia has led to a further charge by some blacks that the government is attempting to use the disease to discourage black births and thereby limit the black population. There has been great controversy over whether or not traits should bear children. Some contend that the couple should be informed of the probable chances of their offspring inheriting the trait or disease, and to offer family planning information only if the people involved requests it. On the other hand, there are those who argue that trait couples should be advised not to bear children. They point out that this would eliminate the possibility of anyone inheriting two genes for the disease, and the disease would then disappear in a century or so. But even

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[1]Smith, "Socio-psychological Aspects," p. 6.

advocates of such a plan are wary of its genocidal overtones.

In addition to the question of whether or not traits should have children is the debate over whether or not persons with the disease should have children. A majority of those who have commented, feel that no one should dictate or force contraception on patients with the disease. But they point out that all offsprings of sickle cell patients are born with the trait and that pregnancy and child birth is particularly dangerous experienced by a woman with sickle cell anemia.

Given these assertions, it is understandable that many blacks are very sensitive to labeling sickle cell anemia as primarily a black disease.

CHAPTER IV

POLITICAL ASPECTS OF SICKLE CELL ANEMIA

Any consideration of the racial implications of sickle cell anemia research and the attendant publicity it has received must include a look at the politics involved. According to Michaelson:

> It is no accident that the current explosion of interest in sickle cell anemia is taking place in the midst of the most significant turbulence in the history of American medicine. Today the failure of this country's health care system is a widely acknowledged, abundantly documented, and massively publicized fact.[1]

Following several years of neglect, the federal government, as well as many private foundations, have been able to show their concern for this health care problem by providing financial support for research, education projects and screening for the disease. In addition, state legislatures have enacted laws to mandate or permit screening and treatment.

Sickle cell anemia as a national health issue received its first significant public support in the 1971 presidential health message.[2]

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[1]Michaelson, "Interesting Pathology," p. 57.

[2]Congressional Record 117 (February 18, 1971), p. 3019.

Reasons for the delayed response to this disease have been discussed and condemned on scientific, educational, and political levels.[1]

Response and Reaction

The response, now that it has come, not only has been inspiring but also, as one commentator has said, "represents progress amid chaos."[2] It is almost as enthusiastic as if the nation were seeking to make up for more than a half-century of neglect by compressing all types of projects and programs into a half-decade of massive effort. In February 1971, President Nixon in his health message 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."[3]

At that time President Nixon announced that he would request six million dollars in congressional funds to combat sickle cell anemia. In May 1972 when Congress passed the National Sickle Cell Anemia Control Act which authorized $115 million to be spent on sickle cell disease over the following fiscal years. The national act provides

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[1] Zueler, "Pediatric Perceptions," p. 340; and Greenberg, "Need to Identify Sickle Cell Trait," p. 630.

[2] "Sickle Cell Disease--Progress Amid Chaos," Medical World News (May 28, 1971):19.

[3] "White House Conference on Children: Report to the President," (Washington, D.C., 1971).

for grants to be made to state and private organizations and agencies for sickle cell programs that would screen and counsel, conduct research, educate the public about the disease.[1] In a matter of two years (1970-1972), the financial picture for sickle cell anemia research and treatment changed drastically.

The storm over funding for sickle cell anemia also extends to money raised by newly-established charity groups and by large-scale benefits. There is little accurate information about the number of sickle cell foundations that existed before 1970, and it is difficult to pinpoint the number that have been established since then. However, every city or state with a sizeable black population appears to have one or more sickle cell fund raising groups.

This display of concern for a predominantly black health problem seemed so sudden and overwhelming that it is now regarded suspiciously. It is felt by many that this sudden interest in sickle cell anemia is politically oriented. For instance, the executive director of one of New York's sickle cell foundations summed up the attitude of many when she said:

> In 1970 the Administration was thinking about the elections of 1972, and looking for a way to get on the good side of blacks. I think the government's interest in sickle cell anemia was clearly politically motivated.[2]

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[1]Public Law 92-294 (92nd Congress, S2676, May 16, 1972).

[2]Cerami and Washington, Sickle Cell Anemia, p. 90.

The Nixon Administration, during its first two years in office, had been criticized by blacks for doing little or nothing in areas that affected the black population. Congress' record was not regarded any more highly than the President's. In the months that passed by Congress, the sickle anemia cause was picked up by politicians around the country. The bill passed Congress unanimously, but those who view the government's concern with sketicism cite the budget cuts of 1972 and 1973 which crippled or eliminated many national programs.[1]

While no one would suggest that the government suspend its sickle cell effort because its motives may not be pure, many blacks feel that if officials want to show blacks they care, they must also enact legislation and allocate funds for comprehensive health care, housing, education, and other areas where help is needed.

Legislative and Legal Support

Eventhough sickle cell disease has been considered chiefly as a medical, research, and educational problem, legal guidelines must be instrumental in providing necessary service and protection to individuals affected, especially children and legal safeguards for those seeking to help in professional and voluntary capacities. In the case of sickle cell anemia the law can and should be considered another signi-

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[1]Ibid., p. 90.

ficant health modality. In this way it can function to help recognize the disease, prevent or limit the occurence and effects of the disease, control spread of the disease, and otherwise assist individuals and their families in communities so that concern for the victims of the disease does not become a factor for discrimination or other adverse consequences. As an adjunct method, however, the law and its strengths must necessarily follow and support the primary approaches; medical, educational, social and also help to provide financial, political, and administrative aid.

Logical sequence suggests that medical and legal priority be given to case finding. By now, screening methods and clinical diagnosis have been established as reasonably effective, safe, and economical when performed by competent staff. States have also passed sickle cell testing laws of which most are voluntary, but according to Bowman, there are at least fifteen states which have compulsory sickle cell testing laws.[1] There are only twenty-two states that have no sickle cell laws.[2] The Virginia law states:

> Every child... shall on or within 30 days after beginning attendance in a public kindergarten, elementary or secondary...furnish a certificate from a duly licensed physician certifying that the child has been tested...

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[1]Bowman, "Mediolegal Social and Ethical Aspects," p. 35.

[2]Powledge, "New Ghetto Hustle," p. 40.

> or that such tests are not, in his professional judgement and discretion, deemed necessary.[1]

The last sentence of this law suggests that white children will be excluded from mandatory testing. This law has added the mandatory sickle cell examination for inmates or correctional and mental institutions.

The Massachusetts law, signed July 19, 1971 reads:

> Every child which the commissioner of public health, by rule or regulation, may determine if susceptible to the disease known as sickle cell trait or sickle cell anemia, shall be required to have a blood test to determine whether or not he has such disease before being admitted to a public school, or in the first year of attendance in a public school...[2]

There are indications that the Massachusetts law has not been fully enforced. In addition, substitute legislation which provide voluntary screening for sickle cell and other genetic disease, flexibility for public health officials in determining what other disease to be added to the list, and proper counseling and treatments have been advanced by some state officials in Massachusetts. Nonetheless, the impact of the compulsory sickle cell law has had a very negative consequence for the black community.

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[1]Commonwealth of Virginia. An Act of amendment of the code of Virginia, adding in Title 32, a Chapter numbered 32-112.10 through 32-112.19, relating generally to the detection and control of sickle cell trait, April 10, 1972.

[2]Commonwealth of Massachusetts, Chapter 491 or Acts and Resolves 1971, S15A.

In order to pass the sickle cell law in Washington, D. C., the City Council had to declare sickle cell a communicable disease.[1] This does not make medical sense--in fact, it adds to the confusion by suggesting that a genetic disorder is the same as an infectious disease. Many of the sickle cell laws require screening tests before marriage and in most cases these laws just amend the regulations for veneral disease.[2] Given the high rate of veneral disease in the black community and the labeling of sickle cell disease or a black disorder, one can imagine the social psychological impact of this problem and how relating these two disorders can easily be interpreted wrongly by racist forces in our society. But the significantly apparent issues have been identified by Powledge when she concluded: "The genetic screening provision of marriage license laws represents the opening wedge for governmental involvement in genetic criteria for procreation."[3]

Furthermore, these laws have had a difficult time dealing with the question of race. It is believed that a compulsory sickle cell law drawn along racial lines might

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[1]District of Columbia City Council, Regulation 72-9, May 3, 1972.

[2]Powledge, "New Ghetto Hustle," p. 45.

[3]Ibid., p. 45.

be unconstitutional. For this reason, many of the legislators use the word black or Negro in the language of the law. However, the New York marriage license law on testing states "each applicant for a marriage license who is not a Caucasian, Indian, or Oriental race."[1] It is worth noting that New York has two sickle cell laws--one for screening school age children and the other for premarital screening. In general, most states have avoided the color question and indirectly the eugenic implications by shifting the decision of who should be screened to state health officials, doctors or marriage license clerks. By not dealing with these problems, the legislators have helped to intensify the conflict around these issues in the community. It is also ironic that many of the compulsory laws were introduced by black legislators.[2] They are experiencing considerable pressures from both the professional and non-professional groups to work for repealing these mandatory screening laws.

In addition, several groups have attempted to establish guidelines for protecting individuals from mass screening programs. For example, the research groups on ethical, social and legal issues in genetic counseling or genetic engineering

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[1]Ibid., p. 46.

[2]Bowman, "Mediolegal Social and Ethical Aspects," p. 35.

[3]"Ethical and Social Issues in Screening for Genetic Disease--A Group Report," The New England Journal of Medicine 286 (May 25, 1972): 1129-1132.

of the Institute of Society and Life has proposed a set of principles and guidelines for the operation of genetic screening programs.[1] These principles include: (1) a need for well planned program objectives; (2) involvement of communities immediately affected screening; (3) provision of equal access; (4) adequate testing procedures; (5) absence of compulsion; (6) a well defined procedure for obtaining informed consent; (7) safeguards for protecting subjects; (8) open access of communities and individuals to program policies; (9) provision for counseling services; (10) understanding the relationship of screening to realizable or potential therapy; and (11) well formulated procedures for protecting the rights of individuals and family privacy.

Black professional groups have also issued specific guidelines. In 1972, there was a National Conference on the Mental Health Aspects of Sickle Cell Anemia at Meharry Medical College in Nashville, Tennessee. The conference was sponsored by the National Institute of Mental Health and the National Center for Family Planning in cooperation with the Department of Psychiatry of the Meharry Medical College. It should be noted that the conference had participants from a variety of disciplines and professions. At the Medical-Legal Aspects and Insurance Program Workshop, the following resolutions were proposed:

> "1. All sickle cell screening should provide for voluntary participation and <u>never</u> should be compulsory; all present mandatory sickle cell screening laws should be repealed.

"2. All screening to be effective should be preceded by adequate education.

"3. All screening and testing should be provided with follow-up counseling by adequately trained counselors for the positives and provide for adequate referral procedures.

"4. No legislation for voluntary screening, counseling or educational programs should be instituted without total funding including provision for treatment of treatment of those unable to pay.

"5. All records must be kept confidential in accordance with the already established doctor-patient relationships. The records must be kept with the primary screening agency and release of all information should only be made with the written informed consent of individual concerned, or his parent or guardian, where applicable.

"6. In view of the fact that persons with sickle cell trait (HbAS) have no disability, they should not be subject to any discrimination.

"7. Mass sickle cell screening by persons, groups or organizations which have no expertise in education, technical aspects of screening or in genetics counseling is an abomination against Afro-Americans and must cease forthwith. If these groups do not do so, they must be made to know that they can be held liable in court for damages.

"8. It is recommended that the Division of Biological Standards of the U. S. Government (or the appropriate agency) withdraw its license from health profiteers which allows the same to market sickle cell tests with misleading, inaccurate, unscientific claims. Presently, electrophoresis is the only practical, primary screening method (there are expensive elaborate research techniques) whereby sickle cell trait (HbAS) may be distinguished from sickle cell disease (HbSS, HbSC, HbS/thalassemia, etc.). To suggest otherwise is criminal and yet another example of commercial exploitation of black people."[1]

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[1] Sickle Cell Screening-Medical-Legal-Ethical, Psychological and Social Problems: A Sickle Cell Crisis, Proceedings, National Conference on the Mental Health Aspects of Sickle Cell Anemia, (Nashville, Tennessee, 1977) (In Press).

The Meharry report, in many instances, seems to be an overreaction to the sickle cell crisis. It also has the problem of how to implement the guidelines. Nonetheless, the report provide some basic frame of reference for mass screening for sickle cell anemia. In addition, these guidelines can be applied to screening programs for other genetic disorders.

Non-Profit Organizations

In the last three years the public has grown increasingly aware of the nature and physiological, sociological and political effects of sickle cell disease. This awareness has led to the establishment of a number of non-profit organizations across the United States to provide information and raise funds for research on the disease. Several of these organizations employ medical personnel and social workers and they offer testing and counseling service or referral for those with the trait or the disease.

There has been a number of federal programs and state and local programs developed in many of the states where there is a sizeable black population. However, as more federal and private funds are made available, and since funds for other social programs as model cities, community action, and day care are drying up, one can expect intensive competition among these groups for these limited resources.[1]

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[1]Black Panthers (May 27, 1972): 11.

There are reports that fly-by-night sickle cell groups have sponsored money raising events, but the funds from the activities seldom reach legitimate sickle cell treatment and research centers.[1] Most state and local governments require a permit to raise funds, but as in many other areas of our society, the law enforcement agencies have not enforced these regulations in the black community.

Moreover, there is the possibility that local volunteer sickle cell groups will not be able to get their share of funds for fighting this disease because they do not have the credentials, influence, tract records, and grantsmanship skills that traditional white oriented institutions have for developing comprehensive programs and proposals for funding. Furthermore, there is the possibility that white organizations will take over local programs and control the sickle cell arena. This would be most unfortunate since this disorder primarily affects the black community in the United States. Blacks should play the key role in the setting of policy decisions in this area. In the meanwhile, however, instead of unity on this common problem, the black community has experienced considerable internal conflict over this health policy issue, thus creating some unanticipated conflictual areas that have a direct bearing on other problems facing the black community.

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[1]Washington Post (November 15, 1972): A12.

CHAPTER V

CONCLUSION

Sickle cell anemia a distinctive malformation of red blood cells, is one of a group of blood disorders inherited from parents. The cellular changes that take place because of this disease may lead to a variety of symptoms that can affect all organ systems of the body. In 1910, the first doctor to publicly announce he had detected an anemia caused by sickling of the blood's red cells was Dr. James Herrick. During the next 40 years, various approaches to research on sickle cell disease were written about, but it was not until 1971, when enough scientific progress had been made, that news of sickle cell disease was reported. Publicity was given to the disease, screening programs were finally developed to detect the disease and groups were organized for funds directed toward research and counseling. In the middle of 1971 the United States Federal Government announced an allotment of $6 million to start special research and other programs on the sickle cell disease. The disease is by now a well-recognized clinical entity that is readily confused with other local and generalized disorders because of its protean characteristics. The disease affects blacks primarily, but it is found in persons and their descendants

from West India, Central and South America, the Middle East, the Mediterranean region and India. It may affect Puerto Ricans and Caucasians who may have inherited the abnormality.

In the United States the outstanding feature of sickle cell disease in terms of social, political, and health implications applies specifically to those with Negroid ancestry. The nature of the disease, the present state of therapeutic impasse, and the increase in the Negro population in many of our large urban centers clearly mark sickle cell anemia as a problem of the gravest public health concern. The disease has been prevalent in the United States for approximately 60 years. After many years of neglect, the federal government, as well as several private foundations, have shown their concern for this health care problem by providing financial support for a research and education projects. However, this sudden interest in the disease has brought about a major controversy in the black population. There are several major factors which underlie this controversy. First, the precedence of this problem relative to other health problems facing the community has been widely argued by black professional and community groups. It is believed by many blacks that too much emphasis on sickle cell disease will divert the community's interest and resources from other health problems such as hypertension, malnutrition, and lead poisoning that have a greater impact on the health development of black people. Secondly, inadequate publicity about the racial and genetic factors of sickle cell anemia has led to a

further charge by some blacks. For example, some of the literature and publicity about the disorder have exaggerated its hazards and classified it as a black man's disorder. The trait and the disease has in many instances been used interchangeably. As a result of these factors there has been panic within some black communities. Thirdly, sickle cell testing laws have been passed in many states sponsored to a great degree by black legislators. These legislators have been ill-advised, and, in too many instances, do not make sense from a medical perspective. Fourthly, the level of internal conflict within the black communities has increased as a result of this issue and many black organizations are fighting each other.

There have been a lot of comments about the political motivation of this newly developed interest in the sickle cell problem. Whatever, the motivation behind this new flow of funds and new surge of interest, it must be satisfying to those who have been frustrated for so long to see that others are finally listening and acting.[1] In regard to the present boom of interest it has been said that:

> All is chaos now, with a lot of researchers running around flag-waving, headline-grabbing, trying to qualify for the therapy and screening jackpot, with the government looking for quick results for high visi-

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[1]C. F. Whitten, "Sickle Cell Programming--An Imperiled Promise," New England Journal of Medicine 288 (1973): 318-324.

> bility in 1972. But it'll fade away, and we blacks will be left with the problem.[1]

It is suggested, however, that this will not happen because public awareness will probably be heightened to such a degree that it will not tolerate neglect of this health care problem and hopefully, some of the new research will hit the target.[2]

Moreover, the health policy issue and others which are directed at the black community must be examined from the perspective of self determination in policy formation. The various sectors of our society must seek control over those programs which are designed to provide specific services to them. In many ways, the attempt on the part of community groups to gain control of the sickle cell health delivery system indicates that there is a crisis in the American medical health "apparatus." However, these groups must not, themselves, fall into the trap of providing second class medicine to the black community in response to the failure of the medical profession. Bowman argued:

> ...no sickle cell program should be instituted unless it can offer competent education, expert technical skills, judicious, professional humane genetic counseling, and adequate service and follow-up. Unless

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[1]Scott, Sickle Cell Anemia and Your Child, p. 271.

[2]Arthur J. Salisbury, "Commentary on Biomedical Research, Sickle Cell Disease, and Community Health," in Sickle Cell Disease, eds. Harold Abramson, John F. Bertles, and Doris L. Wethers (St. Louis, 1973), p. 292.

these goals can be accomplished, our communities are better off with no sickle cell projects.[1]

The black community must view the whole sickle cell anemia issue with some caution. The basic health policy problem facing the black community is the development of mechanisms for controlling a comprehensive health delivery system which includes not only the problem of sickle cell anemia, but also other relevant diseases facing the community.

The legal and ethical factors in consideration of sickle cell disease as a community problem should include the following:

"1. A framework that is based on legislation, case law, and administrative practice. Within such a framework public programs combating this disease can be properly established and managed so that individual and community rights to good health and safe living can be expressed and achieved, using the legal process as one instrument.

"2. Respect for the rights and responsibilities of all participants--those served and those who serve. Concern for the problems of legal liability and the justification of defense of the physician and other associated practitioners must be matched by equal attention to patients who are entitled to be properly informed, protected, and treated.

"3. Recognition and appreciation of community interests--how they are served through the legal system. Individual versus social rights must be balanced; and due process, legal and moral, based on adequate information must be extended to all."[2]

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[1]Bowman, "Mediolegal Social and Ethical Aspects," p.39.

[2]Irving Ladimer, "Legal Considerations in Screening, Treatment, Counseling, and Research in Sickle Cell Disease," in Sickle Cell Disease, eds. Harold Abramson, John F. Bertles, and Doris L. Wethers (St. Louis, 1973), p. 312.

These concepts can prove to be helpful by preventing difficulty, by anticipating problems, planning well in advance, and using the law constructively to head off foreseeable impediments to good practice and good health.

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