# EFFECT OF SICKLE-CELL TRAIT ON REPORTED SYMPTOMS

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ABSTRACT

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ON REPORTED SYMPTOMS

By

Linda M. Walker

The purpose of this study was to determine if there is a difference in reported symptoms between persons with sickle-cell trait. The subjects were Black males and females from the community of Lansing, Michigan. In all, 900 subjects were given blood tests for sickle-cell trait, and 53 were found to have the trait. Each subject completed a questionnaire before the blood test. If a person received notification of a positive test for sickle-cell trait, he came to the Lansing Model Cities office where he filled out the questionnaire a second time and was given genetic counseling.

There was a significant difference between sexes on symptom reporting on both administrations of the health questionnaire. Statistical analysis demonstrated that the main effect of sex ($F=7.88$, $df=1$, $p < .05$) and pre-post test ($F=4.50$, $df=1$, $p < .05$) were significant; in addition, the

interaction sex-pre-post test ($F=4.50$ $df=1$, $p < .05$), and the three-way interaction of sex, trait, pre-post test ($F=4.50$, $df=1$, $p < .05$) were significant.

The above results suggest that sex is related to the amount of symptom reporting, with females reporting more. This was attributed to the fact that women are physically more reactive to illness or stress and this physical sensitivity may condition them to greater awareness of, or concern with, the physical manifestations of illness or stress.

The results also suggest that more information needs to be made available to the public concerning the meaning of sickle-cell trait.

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By

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TO MY MOTHER,

RUBY,

and

ALTON

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# INTRODUCTION

## Need

There is a great need for extensive education of the public about the meaning of sickle-cell disease. Many people have little or no knowledge of the disease, and of those who do, there are many misconceptions. One misconception is the confusion or lack of distinction persons make concerning sickle-cell trait.

An attitudinal survey might enable researchers to find out what people know or don't know concerning this disease. Such research on sickle-cell trait would contribute needed information concerning the knowledge and understanding, or lack thereof, of sickle-cell trait. It is my contention that such information will facilitate the preparation of more and better literature concerning sickle-cell trait and its implications.

Dissemination of literature to medical professions, schools, and paraprofessionals who are in constant interaction with the masses of people will aid in fuller understanding of the disease.

Purpose

Sickle-cell trait (SCT) is more prevalent in some communities than in others. This fact would make one wonder what factors--sex, education, socioeconomic status, age, etc.--are associated with the prevalence of the trait. The purpose of this study is to determine if there is a difference in reported symptoms between persons with SCT. Comparisons of sex, and those with the trait versus those without the trait will be made on the population, to see if any particular factor is associated with a higher incidence of reported symptoms.

It is my belief that there may be a difference in the overall reporting of symptoms between males and females. This difference may not be significant on the first questionnaire; however, after notification of a positive test result for SCT, there will be a significant difference shown upon administration of the same questionnaire a second time. The difference will be in the number and/or severity of reported symptoms.

## REVIEW OF LITERATURE

The disease, sickle-cell anemia (SCA) originated in Africa, where its prevalence is very high in many tribes, particularly in those areas where malaria is endemic. The malaria parasite shuns blood which sickles and it is this property, the greater ability of individuals with sickle-cell disease to survive in these regions of the endemic malaria, which probably accounts for its high incidence. It has been disseminated over all those parts of the world where people of African origin have migrated and intermarried. The occurrence of sickle-cell genes in blood represents not a racial, but an evolutionary mutation that is inherited (Hill, 1970). SCA is now found in certain parts of Africa, in the Mediterranean countries, India and Pakistan, in the Carribbean and in all parts of South, Central, and North Americas. SCA was first recognized by Herrick in 1910. He noticed the phenomenon in a severely anemic West Indian Black student residing in Chicago.

Sickle-cell disease is a noncontagious inherited disorder which occurs in the severe form (the anemia) in one out of every 400 Black Americans, and in the mild form, or trait, in one out of every 10 Black Americans (Francis,

et al., 1970). If two people who have SCT mate, the chances are that one out of four of their children will have the severe form of the disease (i.e., SCA). If one parent has SCA, and the other parent the trait, half of the offsprings will have the trait, the other half the anemia. The life span of individuals with SCA is limited--usually they do not survive beyond their forties. There is no known medical cure.

The clinical manifestations of sickle-cell disease depend primarily upon the high content in every erythrocyte (red blood cell) of the genetically transmitted abnormal hemoglobin S. Sickle-cell hemoglobin is one of many different types of abnormal hemoglobins that occur when one or more of the protein components of normal hemoglobins are changed in their chemical structure. Normal hemoglobin, the oxygen carrying part of the red blood cells, is composed of three chief components: hemoglobin A (HbA), hemoglobin A prime (HbA'), and fetal hemoglobin (HbF) (Hill, 1970). The normal adult hemoglobin (HbA and HbA') can be altered by one or two abnormal hemoglobins. In such a case a person's blood, instead of containing normal hemoglobins, may contain HbA and the abnormal HbS (sickle hemoglobin).

Very few infants with active sickle-cell disease (the anemia) have been described whose symptoms began in the first weeks of life, and not many in the first few months of the first year. This may be due to the high levels of fetal

hemoglobin which in early infancy protects infants from sickling difficulties (Booker, et al., 1964). Not until four months of age, after the adult level of sickle hemoglobin has been attained and the fetal hemoglobin level has begun to decrease, will the red cells begin to sickle in vitro (Booker, et al., 1964).

In environments low in oxygen, hemoglobin S tends to crystallize, causing the cells to sickle. The change in shape (which is reversible) increases the viscosity of the blood and causes local stagnation (slowdown of blood circulation in a part of the body). These processes in turn lead to transitory vascular occlusion and interfere with the circulation in the affected organ. Sickled erythrocytes are mechanically more fragile, are phagocytized (eaten by white cells) more readily, and destroyed more rapidly in the circulating blood than are nonsickled-cells (Diggs, et al., 1933). Therefore, they have a shorter life span, contributing to anemia (lack of red blood cells) and hypoxia (lack of sufficient oxygen). The interaction of these two pathologic processes, anemia plus generalized vascular occlusion, are responsible for the bizarre symptoms and the physical findings associated with SCA (Scott, et al., 1966).

There are basically two groups into which the symptoms and signs of SCA may be separated: (1) those which result mainly from the rapid destruction (hemolysis) or decreased production (bone marrow hypoplasia) of red blood

cells, and (2) those which result mainly from stagnation or poor circulation (Scott, et al., 1966). The former group results in varying degrees of anemia with hypoxia of the body tissues. Patients with SCA tend to compensate adequately for the anemic episodes by the increased production of erythrocytes in the bone marrow. This compensation is thwarted when the marrow is depressed or when the hemoglobin falls to very low levels. Compensation for stagnation is more difficult, especially when this occurs in a vital organ such as the brain, or in any other organ with deficient collateral circulation (Diggs, et al., 1933). Stagnation is more serious in that its effects on organs of the body tend to do more damaging than do those which follow destruction of red blood cells. The following is a detailed outline of complications and symptoms of SCA (Scott, et al., 1966).

Cardiac Enlargement. One of the most common complications of SCA, this expression of dilation and hypertrophy of the myocardium is frequent at all ages. Most patients with SCA tend to have a rapid heart rate. Congestive heart failure may take place at any age and may be related to a previously damaged or impaired mechanism or to repeated pulmonary infections or both.

Respiratory Tract Infection. Infections of the respiratory tract are usually common at all ages. This marked susceptibility is attributed to the combination of long standing anemia, chronic debility, poor nutrition, and inadequate responses of the body's immunologic mechanism. Pneumonia is seen frequently, often developing quickly after a cold; it tends to spread rapidly and to resolve slowly. These patients often require additional support (besides antibodies) of transfusions, in order to elevate the concentration of circulatory hemoglobin to physiologic levels during pulmonary illness. Transfusion treatment seems to accelerate satisfactory recovery.

Central Nervous System. Symptoms and physical signs of neurologic involvement are not uncommon. Symptoms vary from headaches, weakness and lethargy to paralysis, convulsions and coma. They are due either to episodes of cerebral anoxia or to occlusion of the brain blood vessels.

Priapism. This complication is infrequent in young boys, more frequent in adolescents and adults. The painful erection is occasionally of short duration, though it may persist for one to two weeks.

Cutaneous Ulcers. Less common in children than in adults. The lower leg near the ankle is the most favored area.

Retarded Growth and Development. A slowed somatic growth may become recognizable as early as the sixth month of postnatal life. The inadequate development of these individuals is reflected in short stature, thin extremities, long fingers and toes, protuberant abdomens, poor state of nutrition, abnormal posture, hypogonadism, delay in the appearance of secondary sex characteristics, and delay in the onset of menarche. Deceleration in weight gain appears to be associated with frequent infections, increased sickling, and onset of a crisis (Booker, et al., 1964 and Scott, et al., 1966).

A crisis is an alteration that is unpredictable, appears suddenly, and affects a person adversely. In SCA pain crises or simple crises result from the obstruction of the small blood vessels in the bone by the sickled-cells, causing a reduction in the amount of blood normally supplied to the bone and subsequent pain (Robinson, 1970). Swelling of the affected areas accompanies the pain. Crises last for four to six days. Generally during a crisis the pain is severe the first few days only. The swelling usually persists for one to three weeks after the pain has disappeared.

The many complications of the disease not only affect the individual but the family and society as well. From earliest infancy there is the constant medical problem with suffering and recurrent expense. School age children are constantly absent from school because of frequent bouts of pain and chronic fatigue. School achievement is also greatly affected. Physical activity and participation in competitive sports are severely limited. Such restrictions can produce periods of depression in a child and make matters extremely difficult for all concerned.

Persons with SCA must have sedentary vocational employment. Employers are affected by employees with SCA in that the high rate of absenteeism affects work production. It is for this reason and many more that employers and society as a whole must have an understanding of the illness so that working conditions and other situations may be made flexible for those with SCA.

All persons with sickle-cell hemoglobin do not have the severe form of the disease, these people only possess the trait. The sickle-cell trait is typically an asymptomatic condition. A case study of Negro residents of Charleston County, South Carolina, by Boyle, Thompson, and Tyroler in 1960-1964, revealed a high prevalence of the sickle-cell trait at 14.6 percent. The prevalence did not differ between the sexes, nor did it vary significantly with

age groups (ages 35-74). Thus, there is no suggestion based on these cross-sectional data of selective mortality in adults associated with possession of the SCT.

The prevalence of the SCT was significantly higher in the occupational groups of lowest socioeconomic status. Among individuals of low socioeconomic status, the prevalence was higher in residents of rural areas of Charleston County. Probable explanations for these findings include selective malarial mortality in childhood, cultural, and physical isolation, assortive mating, and endogamy (Boyle, et al., 1968). (See Tables 1, 2, 3, and 4.)

Occasionally patients with SCT may develop complications under exceptionally hypoxic conditions such as occur when an airplane with a nonpressurized cabin files at great heights. An instance of complications associated with SCT was reported in the New York Times of February 7, 1970. It stated that four Black servicemen died suddenly as a consequence of vigorous exercise at high altitudes (Hill, 1970; Ragab, et al., 1970). As a result of this incident, a study of one-thousand Black military recruits was performed by Majors R. A. Binder and S. R. Jones of the United States Marine Corps (Binder, et al., 1970). Using the sickledex test the experimenters found that 75 of the one-thousand recruits had hemoglobin type S. The recruits were subjected to the same stress and environment as the aforementioned servicemen who died. No deaths occurred in these recruits.

Table 1. **Distribution of Charleston Negro Men by Occupation and Estimated Income.**

| Occupation | 1960 Census Median Income[8] ($) | 1960 Census No. Employed[8] | Study Sample No. Examined | 1960 Census (%) | Study Sample (%) |
|---|---|---|---|---|---|
| Professional, technical Kindred | 3,174 | 323 | 6 | 2.5 | 1.8 |
| Managers, officials, and proprietors | 1,887 | 194 | 8 | 1.5 | 2.4 |
| Clerical and kindred | 2,508 | 282 | - | - | - |
| Sales workers | 1,115 | 129 | 6 | 3.2 | 1.8 |
| All white-collar | - | 928 | 20 | 7.2 | 6.0 |
| Craftsmen, foremen, and Kindred | 1,973 | 1898 | 49 | 14.9 | 15.0 |
| Operatives and Kindred | 1,891 | 3102 | 45 | 24.3 | 13.8 |
| Service workers | 512 | 1462 | 26 | 11.5 | 8.0 |
| Laborers except farm | 1,465 | 3473 | 147 | 27.2 | 45.0 |
| All blue-collar | - | 9935 | 267 | 77.9 | 81.8 |
| Farmers and farm manager | 709 | 176 | 15 | 1.4 | 4.6 |
| Farm laborers | 644 | 658 | 15 | 5.1 | 4.6 |
| All farm | - | 834 | 30 | 6.5 | 9.2 |
| Occupation not reported | 1,712 | 1078 | 10 | 8.4 | 3.1 |
| All occupations | 1,684 | 12,775 | 327 | 100% | 100% |

Table 2. Distribution of Charleston Negro Women by Occupation and Estimated Income.

| Occupation | 1960 Census Median Income[8] ($) | 1960 Census No. Employed[8] | Study Sample No. Examined | 1960 Census (%) | Study Sample (%) |
|---|---|---|---|---|---|
| Professional, technical Kindred | 2,827 | 777 | 19 | 8.0 | 6.2 |
| Managers, officials, and proprietors | 926 | 84 | 7 | 0.9 | 2.3 |
| Clerical and kindred | 1,219 | 145 | - | - | - |
| Sales workers | 753 | 74 | 3 | 2.2 | 1.0 |
| All white-collar | - | 1080 | 29 | 11.1 | 9.5 |
| Craftsmen, foremen, and Kindred | 1,973 | 35 | 1 | 0.4 | 0.3 |
| Operatives and kindred | 1,064 | 900 | 26 | 9.3 | 8.5 |
| Service workers | 541 | 6218 | 34 | 64.0 | 11.1 |
| Laborers except farm | 812 | 51 | 162 | 0.5 | 53.1 |
| All blue-collar | - | 7204 | 223 | 74.2 | 73.0 |
| Farmers and farm manager | 571 | 25 | - | 0.3 | - |
| Farm laborers | 511 | 660 | 45 | 6.8 | 14.8 |
| Occupation not reported | 712 | 735 | 8 | 7.6 | 2.6 |
| | - | 9704 | 305 | 100.0% | 100.0% |
| Housewives | - | - | 143 | - | - |
| All occupations | 650 | - | 448 | - | - |

Table 3. Prevalence of the Sickle Cell Trait by Age and Sex*

| Age | Men | | Women | | Both Sexes | | | | |
|---|---|---|---|---|---|---|---|---|---|
| | No. Tested | No. S.C.T. | No. Tested | No. S.C.T. | No. Tested | No. S.C.T. | Men | Women | Both Sexes |
| 35-44 | 142 | 22 | 193 | 27 | 335 | 49 | 15.5 | 14.0 | 14.6 |
| 45-54 | 79 | 8 | 104 | 16 | 183 | 24 | 10.1 | 15.4 | 13.1 |
| 55-64 | 64 | 11 | 76 | 11 | 140 | 22 | 17.2 | 14.5 | 15.7 |
| 65+ | 42 | 7 | 75 | 11 | 117 | 18 | 16.7 | 14.7 | 15.4 |
| All Ages | 327 | 48 | 448 | 65 | 775 | 113 | 14.7 | 14.5 | 14.6 |

*Charleston, SC.

Table 4. Prevalence of the Sickle Cell Trait by Occupations.*

| | Men | | Women | | Both Sexes | | |
|---|---|---|---|---|---|---|---|
| | No. Tested | No. with S.C.T. | No. Tested | No. with S.C.T. | No. Tested | No. with S.C.T. | Both Sexes % with S.C.T. |
| Upper social class sample | 103 | 6 | - | - | 103 | 6 | 5.8 |
| Probability sample: | | | | | | | |
| Occupation | | | | | | | |
| White-collar | | | | | | | |
| Professional, technical, and Kindred | 6 | 0 | 19 | 1 | 35 | 1 | 2.9 |
| Proprietors and managers | 8 | 1 | 7 | 1 | 15 | 2 | (13.3) |
| Clerical and sales | 6 | 1 | 3 | 1 | 9 | 2 | (22.2) |
| Blue-collar | | | | | | | |
| Craftsmen and foremen | 49 | 5 | 1 | 0 | 50 | 5 | 10.0 |
| Operatives and kindred | 45 | 8 | 26 | 2 | 71 | 10 | 14.1 |
| Protective and service | 26 | 3 | 34 | 3 | 60 | 6 | 10.0 |
| Labor, nonfarm | 147 | 26 | 162 | 30 | 309 | 56 | 18.1 |
| Farm | | | | | | | |
| Farm managers | 15 | 2 | 0 | 0 | 15 | 2 | (13.3) |
| Farm laborers | 15 | 0 | 45 | 8 | 60 | 8 | 13.3 |
| Occupations not reported | 10 | 2 | 8 | 2 | 13 | 4 | (22.2) |
| Major occupation groupings | | | | | | | |
| Upper-social class sample | 103 | 6 | - | - | 103 | 6 | 5.8 |
| All white collar | 20 | 2 | 29 | 3 | 49 | 5 | 10.2 |
| All blue collar | 267 | 42 | 223 | 35 | 490 | 77 | 15.7 |
| All farm | 30 | 2 | 45 | 8 | 75 | 10 | 13.3 |
| Occupations not reported | 10 | 2 | 8 | 2 | 18 | 4 | (22.2) |
| All housewives | - | - | 143 | 17 | 143 | 17 | 11.9 |

*Negro, Charleston County, SC.

This study points up the necessity for further study to enable the medical profession to understand more about the complications of and the mechanisms of sickling. Another important point is that of the 75 recruits with hemoglobin S, only five knew of any sickle-cell disease in their family, and there was a complete lack of awareness of it in themselves. Those who were sickle-cell negative did not have any familiarity with the disease entity. This may have been due to the fact that many of the recruits were from rural areas. Whatever the reason, this study shows the need for more education of all people about this disease.

Ragab et al. (July, 1970) did a case study on a six-month old Negro boy who was treated in a hospital for fever, dehydration, severe diarrhea, vomiting and extensive bilateral pneumonia. Blood analysis showed he had SCT. Because of the infection and other illnesses (severe pneumonia, metabolic acidosis) which were present, the sickling phenomenon was precipitated. The resulting sickling led to focal obstruction of the blood supply and thus, left him partially blind. Much of the sickling was brought under control, but had it not and the sickling had become widespread throughout his body blocking vital organs or causing the blood-forming organs to stop producing new blood cells, it may have been fatal.

The last case study points to the fact that under extreme conditions persons with SCT may begin to sickle like

a person with SCA when there are complicating influences from other illnesses. Therefore, trait identification is a vital information in an individual's medical history. Though SCT is typically asymptomatic, such information is valuable in the treatment of patients in areas where there is a relatively high incidence of the trait. Such information will facilitate in the treating of patients without the sudden appearance of unknown factors to complicate matters.

The Foundation for Research and Education in Sickle-Cell Disease has compiled a list of guidelines necessary for education and research in sickle-cell disease (Francis, et al., 1970):

> A) Many doctors are unaware of the disease. Sometimes patients with SCA are treated for diseases which have similar symptoms. More information is needed on SCA at physicians' conventions, dissemination of literature, composition and distribution of a practical office sickle-cell testing kit. Diagnostic test for sickle-cell disease should be a regular part of diagnostic procedures.
>
> B) Teachers need to know what to expect if there is a child with SCA in the class.
>
> C) Guidance and vocational counselors need to know how to direct sicklers into suitable environments.
>
> D) Improved medical procedures will enable the patients to live a fuller and probably longer life.
>
> E) Premarital genetic counseling is greatly needed. This counseling will enable parents to better understand the possible genetic make-up of their offsprings.
>
> F) All health agencies should know the incidence and forms of this disease in the population they serve so that better health programs can be implemented.
>
> G) Much basic hematological research is required because the phenomenon of sickling is only partially known.

H) A list of effects of specific drugs on the illness needs to be compiled. The proper methods of handling transfusion in SCA patients is yet to be fully determined.

## Discussion of Previous Research

Most research thus far on sickle-cell disease has been on the anemia. Research on the trait has been very limited. Of the research done on the trait there is no conclusive evidence that the trait alone causes serious illness, or if there are other existing complicating factors which may influence the sickling mechanism and thereby produce the resulting situation. Each case study does, however, show the need for more research and education on SCT. My paper is directed towards pointing out the necessity for more extensive educational literature.

## Present Study

In this study a random sample of subjects who come to a sickle-cell anemia screening test area will be obtained. A questionnaire will be given to each subject to be filled out before their blood is drawn. Each subject will be notified by mail of their test results (either they will possess the sickling trait or not--SCT positive or SCT nonpositive). Those subjects with a positive SCT result will come to the Model Cities Health Clinic in Lansing for genetic counseling.

Before each counseling session, a second questionnaire will be administered to the subjects (the same questionnaire as used on the pretest).

The pretest responses of those subjects with SCT and those without SCT, will be compared. Also, the change in reported symptomatology after notification of test results will also be investigated.

A psychologist would be interested in this study because of possible emotional disturbances which may occur in subjects who are notified of a positive test result for SCT. These disturbances stem from a lack of understanding and misconceptions concerning the meaning of SCT. The reported symptoms of the subjects on the second questionnaire will show the effects of any possible negative views (i.e., more frequency of reported symptoms) the subject may now hold.

From a psychological viewpoint I feel that the subjects with SCT will expect or anticipate certain conditions they think are associated with SCA and will report that they possess these conditions now, or if possessed before, they are more prevalent now. These expectancies may produce such negative behaviors as depression, which is a behavior of interest to a psychologist.

## DESIGN OF THE STUDY

### Methodology

This is a predictive field study. The design of the study includes a pretest-post test measure with a matching control group.* In the pretest all subjects received a 24-item questionnaire (see Figure 1) which contained symptoms typical of SCA, and which was filled out before receiving the blood test.**

In the post test, the same questionnaire was filled out again by those subjects who received notice of a positive test result for SCT.

The pretest questionnaire was given to 907 subjects, 53 of whom had the trait. A control group of equal number of those without the trait was randomly selected. The number of males and females was the same for both groups.

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*Post test control group was paid $2 per subject for participation. Funding was provided by a grant to Dr. Lawrence Messe from the United States Air Force Research.

**Blood tests were given by the Department of Human Development of Michigan State University. Funding provided by grants from the City Demonstration Agency and the Center for Urban Affairs, Michigan State University.

Figure 1. Health Survey and History.

A. Name _____

B. Address _____

C. Telephone Number _____

D. Age _____ Sex _____

E. Please list everyone in your family who lives with you:

| Name | Sex | Age | Relationship |
|---|---|---|---|
| | | | |

F. Would you say that your health is excellent, good, fair, or poor? (circle one)

G. In the past year have you had trouble with any of the following conditions or diseases? (circle response)

| | | | | | |
|---|---|---|---|---|---|
| 1. Jaundice? (yellow color in eyes or skin) | Never | Sometimes | Frequently | Almost always | Always |
| 2. High or low blood pressure? | Never | Sometimes | Frequently | Almost always | Always |
| 3. Blood in bowel movement? | Never | Sometimes | Frequently | Almost always | Always |
| 4. Blood in urine? | Never | Sometimes | Frequently | Almost always | Always |
| 5. Have you spit blood in the last year? | Never | Sometimes | Frequently | Almost always | Always |
| 6. Nose bleeds? | Never | Sometimes | Frequently | Almost always | Always |
| 7. Faintness or dizziness? | Never | Sometimes | Frequently | Almost always | Always |
| 8. Shortness of breath or trouble breathing? | Never | Sometimes | Frequently | Almost always | Always |
| 9. Tire easily or always tired? | Never | Sometimes | Frequently | Almost always | Always |
| 10. Pain in chest? | Never | Sometimes | Frequently | Almost always | Always |
| 11. Convulsions? | Never | Sometimes | Frequently | Almost always | Always |
| 12. Sore throat? | Never | Sometimes | Frequently | Almost always | Always |
| 13. Swelling or painful joints? | Never | Sometimes | Frequently | Almost always | Always |

Figure 1. Continued.

| | | | | | |
|---|---|---|---|---|---|
| 14. Muscle pain? | Never | Sometimes | Frequently | Almost always | Always |
| 15. Weakness or numbness in legs? | Never | Sometimes | Frequently | Almost always | Always |
| 16. Pain in bones? | Never | Sometimes | Frequently | Almost always | Always |
| 17. Kidney or bladder trouble? | Never | Sometimes | Frequently | Almost always | Always |
| 18. Liver trouble? | Never | Sometimes | Frequently | Almost always | Always |
| 19. Increased urine flow? | Never | Sometimes | Frequently | Almost always | Always |
| 20. Pain in stomach or abdomen? | Never | Sometimes | Frequently | Almost always | Always |
| 21. Unusual growth or tumor on or under the skin? | Never | Sometimes | Frequently | Almost always | Always |
| 22. Ulcers (sores)?--Leg, foot or arm? | Never | Sometimes | Frequently | Almost always | Always |
| 23. Earaches? | Never | Sometimes | Frequently | Almost always | Always |
| 24. Loose bowels? | Never | Sometimes | Frequently | Almost always | Always |

Of those having the trait, only 39 females and four males could be contacted for the post test measure. A randomly selected group of those without the trait served as the control, both groups being matched on sex.

## RESULTS AND DISCUSSION

### Results

A principal axis factor analysis of the 24-item questionnaire with varimax rotation and an eigenvalue cut-off of 1.00 was done. This analysis resulted in four factors. Tables 5 and 6 show the rotated factor matrix with items.

A separate analysis of variance on the summed scores of the items constituting each factor was then performed. The design of each analysis inspected the effects of trait (presence-absence), sex, and measurement sessions (pretest-post test). Factor 1 alone demonstrated significant results (see Tables 7 and 8). The main effect of sex was significant ($F=7.88$, 1,53 df, $p < .05$); and pretest-post test ($F=4.50$, 1,53 df, $p < .05$), and the three-way interaction of sex, trait, pre-post test ($F=4.50$, 1,53 df, $p < .05$) were significant. The interactions above are listed as AC and ABC in the analysis of variance Table 7.

Given the significant three-way interaction it is more appropriate to discuss the interactive effects of the variables under study rather than the significant main effects. Graphic representations of this interaction are shown in Figure 3.

Table 5. Rotated Factor Matrix.

| | Factor 1 | Factor 2 | Factor 3 | Factor 4 |
|---|---|---|---|---|
| 1. Jaundice | -.0419 | -.1641 | .0099 | .4068* |
| 2. High or low blood pressure | .4097* | .0270 | .2548 | -.0635 |
| 3. Blood in bowel movement | .1209 | -.2591 | .3332* | .2027 |
| 4. Blood in urine | -.0537 | .0603 | .6136* | .1053 |
| 5. Have you spit blood in the last year | -.0393 | -.0294 | .1133 | .3357* |
| 6. Nose bleeds | .0078 | -.3920* | -.0415 | .1065 |
| 7. Faintness or dizziness | .5131* | -.1587 | .1827 | -.0955 |
| 8. Shortness of breath or trouble | .6687* | -.0023 | .0765 | -.0729 |
| 9. Tire easily or always tired | .6440* | -.2244 | .1192 | .1521 |
| 10. Pain in chest | .5498* | -.2108 | .0930 | -.0934 |
| 11. Convulsions | .0324 | .0830 | .0844 | .4597* |
| 12. Sore throat | .1222 | -.5603* | -.0067 | -.0358 |
| 13. Swelling or painful joints | .5691* | .0825 | -.0084 | .3667 |
| 14. Muscle pain | .6786* | -.1122 | -.0528 | .3041 |
| 15. Weakness or numbness in legs | .6333* | -.1060 | .1405 | .2382 |
| 16. Pain in bones | .3308* | -.1320 | .1302 | -.3124 |
| 17. Kidney or bladder trouble | .2497 | .0271 | .6888* | .0796 |
| 18. Liver trouble | .0899 | -.1750 | .3060 | .3190* |
| 19. Increased urine flow | .2497 | -.1078 | .5380* | .0314 |

Table 5. Continued.

| | Factor 1 | Factor 2 | Factor 3 | Factor 4 |
|---|---|---|---|---|
| 20. Pain in stomach or abdomen | .2858 | -.4427* | .0742 | .1524 |
| 21. Unusual growth or tumor on or under the skin | .4015 | -.0942 | -.1514 | .5221* |
| 22. Ulcers (sores)--leg, foot, or arm | .1407 | -.1797 | .0778 | .2715* |
| 23. Earaches | .1336 | -.5967* | .0209 | .0804 |
| 24. Loose bowels | .0147 | -.5679* | .1440 | .0556 |
| 25. Hi. Load. | .6786 | -.5967 | .6888 | .5221 |
| 26. Prop. Var. | .2228 | .1134 | .1060 | .0983 |
| 27. Cum. P.V. | .2228 | .3362 | .4421 | .5404 |

Table 6. Factored Items.

| Factor 1 | Factor 2 | Factor 3 | Factor 4 |
|---|---|---|---|
| High or low blood pressure | Nosebleeds | Blood in bowel movement | Jaundice |
| Faintness or dizziness | Sore throat | Blood in urine | Have you spit blood in the last year |
| Shortness of breath or trouble breathing | Pain in stomach or abdomen | Kidney or bladder trouble* | Convulsions |
| Tire easily or always tired | Earaches* | Increased urine flow | Unusual growth or tumor on or under the skin* |
| Pain in chest | Loose bowels | | Ulcers |
| Swelling or painful joints | | | |
| Muscle pain* | | | |
| Weakness or numbness in legs | | | |
| Pain in bones | | | |

*Highest reported symptom in each factor.

Table 7. Analysis of Variance Using Unweighted Means-Factor 1.

| Source of Variation | SS | df | MS | F |
|---|---|---|---|---|
| A (SEX) | 1.89 | 1 | 1.89 | 7.88* |
| B (TRAIT-NON TRAIT) | 0.00 | 1 | 0.00 | 0.00 |
| AB | .09 | 1 | .09 | .38 |
| ERROR 1 | 12.82 | 53 | .24 | |
| C (PRE-POST TEST) | .09 | 1 | .09 | 4.50* |
| AC | .09 | 1 | .09 | 4.50* |
| BC | 0.00 | 1 | 0.00 | 0.00 |
| ABC | .09 | 1 | .09 | 4.50* |
| ERROR 2 | 1.06 | 53 | .02 | |

*Significant at $p < .05$

Table 8. ABC': Cell Means.

| | | Factor 1 | | | |
|---|---|---|---|---|---|
| | | C1 | | C2 | |
| | | B1 | B2 | B1 | B2 |
| Males | A1 | 1.09 | 1.08 | 1.09 | 1.03 |
| Females | A2 | 1.30 | 1.37 | 1.41 | 1.53 |

Legend:
C1 = PRETEST
C2 = POST TEST
B1 = CONTROL
B2 = TRAIT
A1 = MALES
A2 = FEMALES

## Discussion

The factor analysis of the twenty-four item questionnaire resulted in four factors (see Table 7). Going from factor 1 to factor 4, the symptoms increase in their severity (e.g., dizziness - factor 1, is less severe than convulsions - factor 4). Factor 1 consists mainly of everyday complaints which people in general tend to have. These symptoms are generally had by most people at one time or another. Muscle pain was the symptom most often reported. This may be due to the fact that both adults and children at one time or another probably suffered simple muscle pain due to strain or overexertion.

Factor 2 symptoms are a little more serious than factor 1, in that they are less general and usually associated with some mild illness involving pain. Earaches were the most commonly reported, probably because they cause more pain than the other factor 2 symptoms.

In factor 3 are found symptoms associated with urinary tract infection, with kidney or bladder trouble being reported most often. These symptoms are of a more serious nature than those of the first two factors and are indicative of internal malfunctioning.

Factor 4 consists of highly serious and very severe illnesses which may leave one who has them temporarily incapacitated. Whereas symptoms in both factors 1 and 2 may not need a doctor's care, almost all of those in factor 3

do, and all of those in factor 4 definitely require doctor's care. The most frequently reported symptom in factor 4 was unusual growth or tumor on or under the skin. The reason for this symptom being most frequently reported is that it may have been interpreted to mean any minor growths such as bumps from a rash, when in fact it was meant to mean major unusual growths. Therefore, more people would report this symptom.

The experimental design allowed for the random selection of control group subjects. The results of data analysis using this design are graphically shown in Figures 2 and 3. Figure 2 shows mean responses for the trait group (both males and females) on the pretest and post test measures. Females significantly reported more symptoms than males on both the pretest and post test. The higher incidence of symptom reporting by the females may be due to the fact that women readily report their illnesses. Females significantly increased in symptom reporting on the post test, possibly due to lack of understanding of SCT. Typically SCT is asymptomatic. The possible logic behind the increase is that because this group of females may have equated SCT as being the same as SCA they believed they should be ill or should have more symptoms than they previously reported.

Males reported far fewer symptoms on the pretest than did females. A possible explanation for the low number

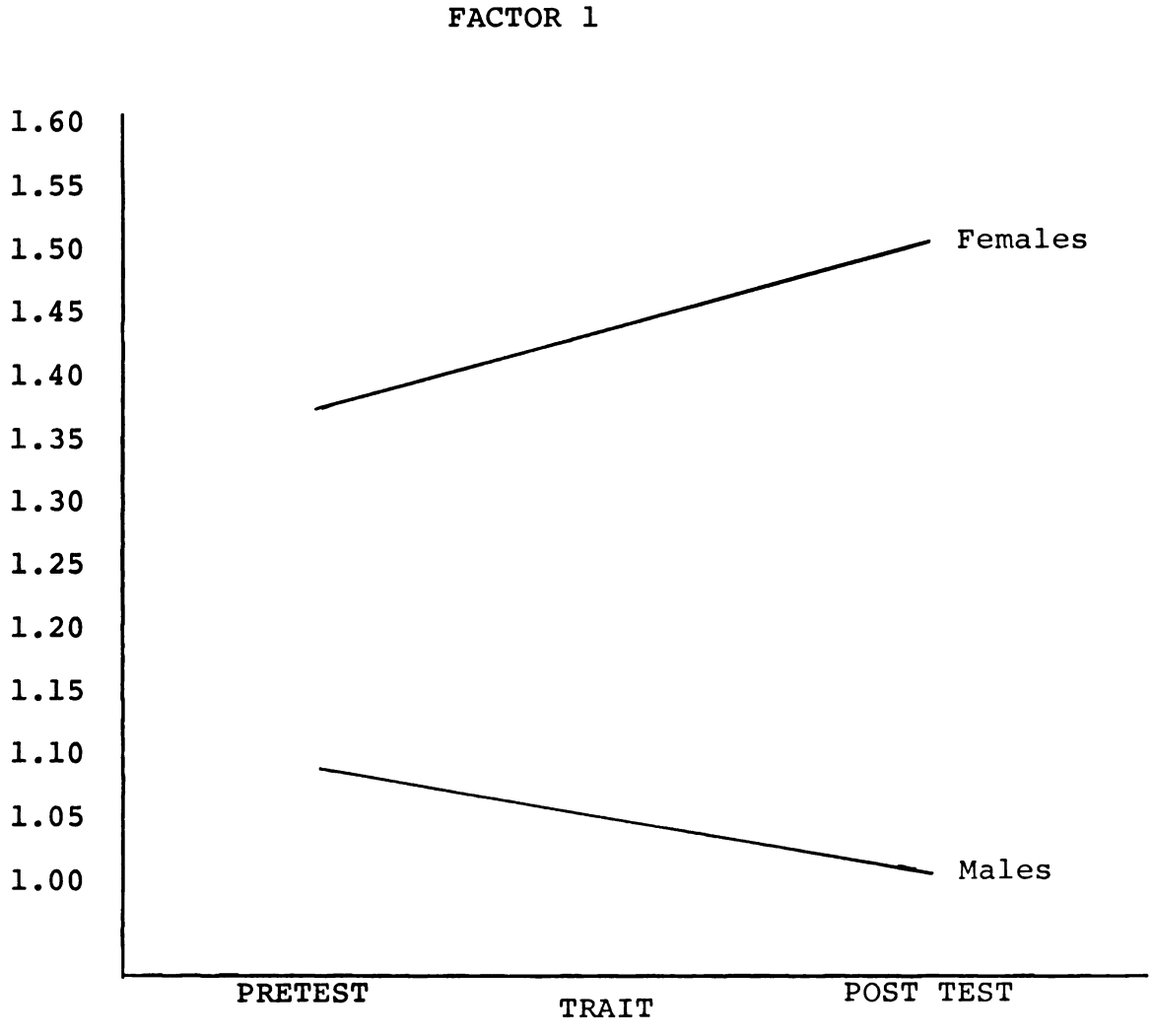


Figure 2. Trait- AC Interaction: Sex, Pre and Post Test.

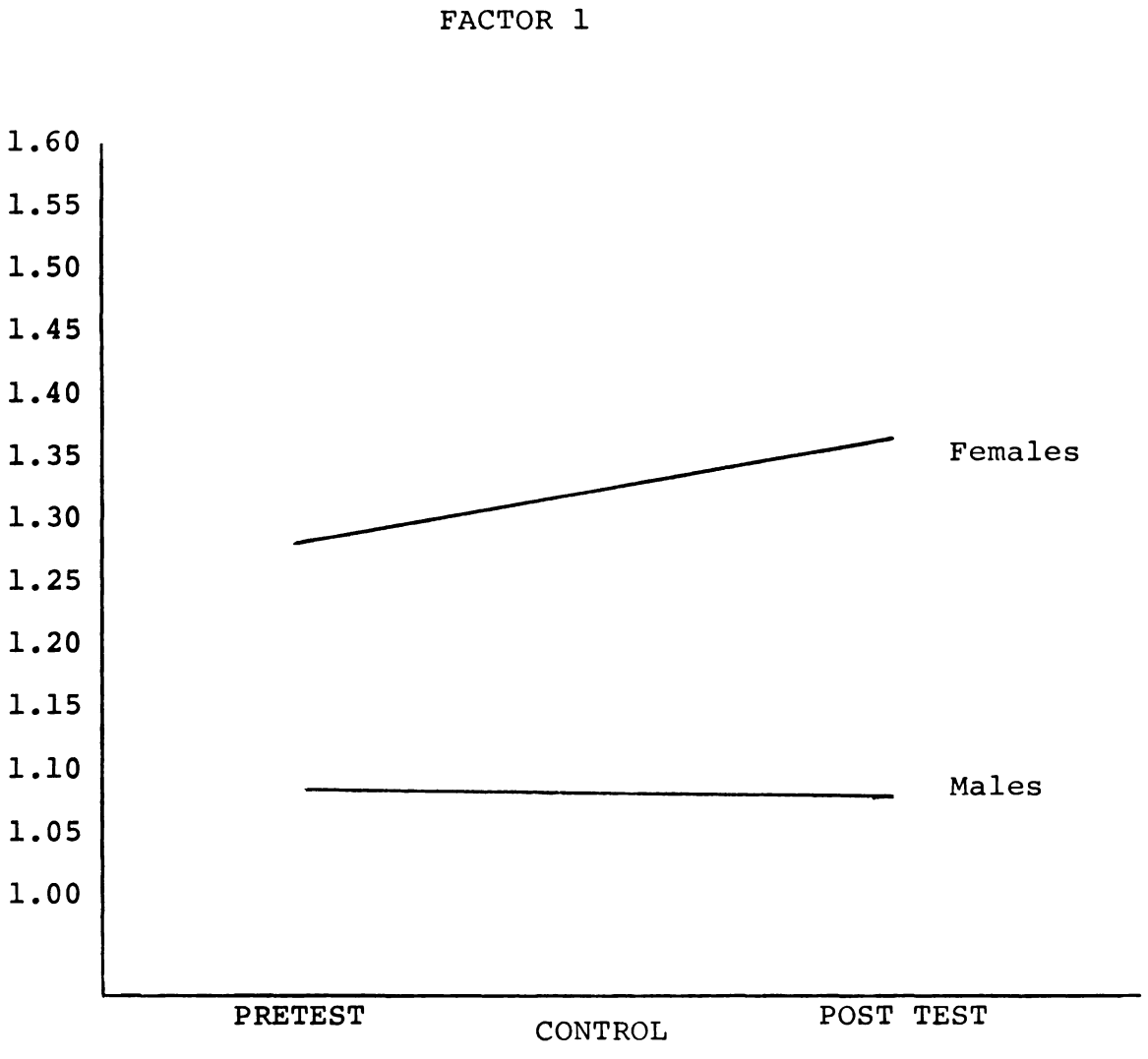


Figure 3. Control- AC Interaction: Sex, Pre and Post Test.

of responses on the pretest is that men are less likely to report or acknowledge their illnesses.

The amount of symptom reporting by males on the post test slightly decreased from that of the pretest measure. This decrease in symptom reporting could possibly be interpreted as a defense by the males to retain their strong masculine demeanor. In our society today males are stereotyped as being strong and healthy and to acknowledge illness (i.e., report more symptoms) would somehow lessen this image.

Figure 3 graphically represents mean responses of females and males in the control groups. The female control group reported many symptoms on the pretest, and even more on the post test. The rationale behind the high incidence of symptom reporting in this group is the same as for the female trait group, namely, more willingness to report illnesses than men. The increase in symptom reporting from pretest to post test may be due to a feeling of safety now that they know they do not have SCT. On the pretest the female control group as well as the trait group may have felt threatened in that they may have believed that the more symptoms they had, the higher their chances of possessing SCT. But, when the control group learned they did not have the trait, the pressure was lifted and they were freer to report more symptoms.

In comparing both female groups, one finds that the trait group reported more symptoms than the control group

when in actuality there should have been no significant difference.

The male control group was low in reporting symptoms, remaining the same on both pre and post tests. A possible explanation being the same as for the male trait group, namely, less likeliness of males to report or acknowledge illness. When the control group received notification that they did not have the trait, they felt no need to report more symptoms. Unlike the females, they were not threatened by the number of symptoms they reported on the pretest, and therefore did not experience the feeling of now being safe to report more symptoms.

Generally speaking, females reported more symptoms in both groups and under both pre and post tests than did the two male groups, all of which may be due to the fact that women are physically more reactive to illness or stress and this physical sensitivity may then condition them to greater awareness of, or concern with, the physical manifestations of illness or stress.

Four analyses of variance were done. Only factor 1 proved to be significant (see Table 7). Of the four factors, factor 1 accounted for the most variance with a wider range of symptoms. The symptoms of factor 1 are commonly had by most people, whereas those of the other three factors are either very severe and not had by most people, or the cluster they form is more specific to a particular part of the body and not very prevalent in the population sampled.

The number of persons with the trait who came to the Model Cities Clinic for genetic counseling was very low (13 out of 53). One reason for the small turn out may be that persons with the trait feared that something really may have been wrong with them so they did not come in. It is possible that males used denial of the trait and therefore they did not come in for the counseling, whereas females did not come possibly out of fear. Females who filled out the questionnaire voiced their reluctance and fear of finding out if they had the trait or not. Those females notified of a positive test result for SCT, and not understanding its meaning, were frightened even more, now that they knew they had the disease. It may be because of this heightened fear that they definitely refused to come in for the counseling.

In summation, there was reluctance on the part of both sexes to come in for genetic counseling. This fact may be attributed to lack of understanding of SCT.

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# SUMMARY AND CONCLUSION

## Summary

During the last few years renewed interest in SCA and SCT has appeared in the United States. This study concerns itself with the reporting of symptoms in a SCT survey on the Black population in the city of Lansing.

A health survey questionnaire was devised by Mrs. Frankie Brown, Mr. Astrid Mack, and Dr. James Higgins of the genetics laboratory at Michigan State University, which facilitated obtaining medical history information from persons coming to a mobile unit to receive a blood test for SCT. The questionnaire contained twenty-four symptoms associated with SCA. A factor analysis of the responses to this questionnaire extracted four orthogonal factors; the items which loaded on each factor were then inspected in four separate analyses of variance.

Nine-hundred subjects received the blood test for SCT, and of these, 53 had the trait. The majority of those with the trait were female.

Attention was focused on:

A) Sex of subject,
B) Trait or non-trait

C) Responses given on the questionnaire before the blood test, and responses given on the same questionnaire a second time after notification of a positive blood test result for the trait.

## Conclusion

The results obtained from the female and male groups show a significant difference in the reporting of symptoms, with females definitely reporting more. Also, because of the significant rise in symptom reporting by the female trait group when SCT is typically asymptomatic lends credence to the belief that the SCT condition is not understood. More research utilizing a larger trait group should show this finding to be very prevalent in the population tested.

# LIST OF REFERENCES

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## LIST OF REFERENCES


Binder, Richard A. and Jones, Stephen R., Prevalence and Awareness of Sickle-Cell Hemoglobin in a Military Population, Journal of the American Medical Association, 214:909-911, November 2, 1970.

Booker, Clifford R., Ferguson, Angella D., Scott, Roland B., Studies in Sickle-Cell Anemia, XXII, Clinical Manifestations of Sickle-Cell Anemia During the First Two Years of Life, Clinical Pediatrics (Philadelphia), 3:111-5, February 1964.

Boyle, Edwin, Jr., Thompson, Charles, Tyroler, Herman A., Prevalence of the Sickle-Cell Trait in Adults of Charleston County, South Carolina, An Epidemiological Study, Archives of Environmental Health (Chicago) 17:891-8, December 1968.

Diggs, L. W., Ahmann, C. F., Bibb, J., Incidence and Significance of Sickle-Cell Trait, Annals of Internal Medicine, 7:769-771, 1933.

Diggs, L. W., Sickle-Cell Crisis, American Journal of Clinical Pathology, 44:1-19, 1965.

Francis, Y. F., Wethers, D. L., Fenwick, L. A., The Foundation for Research and Education in Sickle-Cell Disease, Journal of the National Medical Association, 62:200-3, May 1970.

Hill, George W., Sickle-Cell Anemia: Facts and Fallacies, Essence Magazine, July 1970.

Ragab, Abdel-Salam H., Stein, Mervyn R., Vietti, Theresa J., Severe Complications in an Infant Due to Sickle-Cell Trait, Clinical Pediatrics (Philadelphia), 9: 416-9, July 1970.

Robinson, Lawrence D., Sickle-Cell Anemia-Danger to Our Children, 1970.

Scott, Roland B., Ferguson, Angella D., Studies in Infants and Children in the United States, Clinical Pediatrics (Philadelphia), 5:403-9, July 1966.