CONTRACEPTIVE PRACTICES AND REPRODUCTIVE PATTERNS IN SICKLE CELL DISEASE

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A questionnaire was administered to 52 female subjects with sickle cell disease of genotypes Hgb SS, SC, and S-thalassemia, and to 80 control subjects. They answered questions pertaining to their contraceptive habits, their reproductive patterns, and their sexual activity.

It was found that sexual activity differed significantly for the two groups; only 38 percent of the females in the sickle cell group reported sexual activity compared with 81 percent of the females in the control group. Contraception was used less frequently by the sickle cell group (33 percent vs 66 percent). The most commonly used method was an oral contraceptive. However, barrier methods were chosen by the cumulative majority. The two groups exhibited similar rates of conception but differences in the outcome of the pregnancies: the sickle cell patients experienced more miscarriages and premature births. There was a greater percentage of cesarean sections in sickle cell patients (46 percent) compared with controls (18 percent).

The influence of the sickle cell gene has frequently been evaluated in terms of its effect upon growth and development as well as on pregnancy and its outcome.[1,2] Reports indicate an increased incidence of fetal wastage, prematurity and maternal morbidity in sickle cell patients.[1-3] A relative subfertility in patients with homozygous sickle cell disease (SS) has been documented.[1-4] In the heterozygous state, ie, sickle cell trait, no effect on fertility has been noted.[5] Hendrickse et al reported no evidence of reduced fertility in other sickle hemoglobinopathies such as sickle cell hemoglobin C disease (Hgb S/C) and sickle cell beta thalassemia (Sβ-thal).[2] Delays in growth and development, sexual maturation, and secondary sexual characteristics are well documented.[6-8]

However, there are few available data on the actual contraceptive methods chosen by women with sickle cell disease. Only transient mention of the contraceptive habits of these patients has been noted.[4,9] The purpose of this study is to review preceding research and to address issues that have not been previously reported.

MATERIALS AND METHODS

The study group consisted of female patients with sickle cell disease who attended the Center for Sickle Cell Disease at Howard University. Six patients at the George Washington University hematology clinic who were questioned by the same interviewer were also included. These subjects had Hgbs SS, SC, and S-thalassemia and the control of Hgb AA and sickle cell trait (AS). The hemoglobin pattern of all patients was verified using hemoglobin electrophoresis. Questionnaires were administered to 52 patients, 34 with SS Hgb, 10 with SC Hgb, and 4 with Sβ-thal. Statistics on four of the 52 patients were deleted from many of the analyses because of

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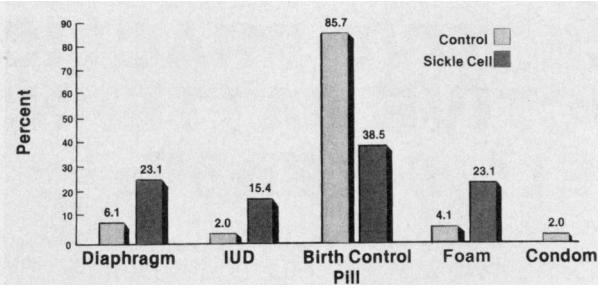

Figure 1. Types of contraception

incomplete questionnaires. The age range of the sickle cell sample was 14 to 46 years (mean, 25.3 years). The control group was composed of 80 patients who attended the Family Planning Clinic at Howard University Hospital. The age range of the control group was 16 to 40 years (mean, 23.7 years). Eighteen percent of the control group were hemoglobin AS.[2,7] Both groups were asked questions pertaining to their menses. The age at first sexual encounter was sought, as well as information on contraceptive habits, eg, the age at which the contraceptive method was first used and the type of method used during the most recent sexual encounter. Data on the outcome of pregnancy, the method of delivery, and the number of unplanned pregnancies were sought. The ages of the oldest and the youngest children were obtained in addition to the number of living children. Demographic data such as marital status were also requested. Subjects were interviewed before and after they completed the questionnaire to answer any questions that needed further clarification.

RESULTS

Sexual Activity

Thirty-nine percent of the sickle cell group reported that they were sexually active compared to 81 percent of the control group ($P < .001$). The mean ages of the first sexual relationship in the sickle cell group and controls were 17.7 years and 17.0 years, respectively.

Contraception

Only 33 percent of the study group reported use of a contraceptive method compared with 66 percent of the controls. Of further significance, only 30 percent of the sickle cell patients had used any contraception during their most recent sexual encounter whereas 64 percent of the controls reported contraceptive coverage ($P < .003$) during that encounter.

The most frequently used method in both groups was the birth control pill (86 percent use in the controls and 39 percent in the sickle cell group). However, it was clear that the sickle cell group used a greater variety of contraceptive methods, with the cumulative majority choosing the diaphragm (23 percent), intrauterine device (15.4 percent), and foam (23 percent). Thirty-eight percent of the sickle cell group reported unplanned pregnancies, which was not a significant finding ($P < .4$) when compared with 49 percent of the controls. Fifty-five percent of the sickle cell group reported that they were using no contraception at that time vs a 35 percent lack of use among the controls (Figure 1).

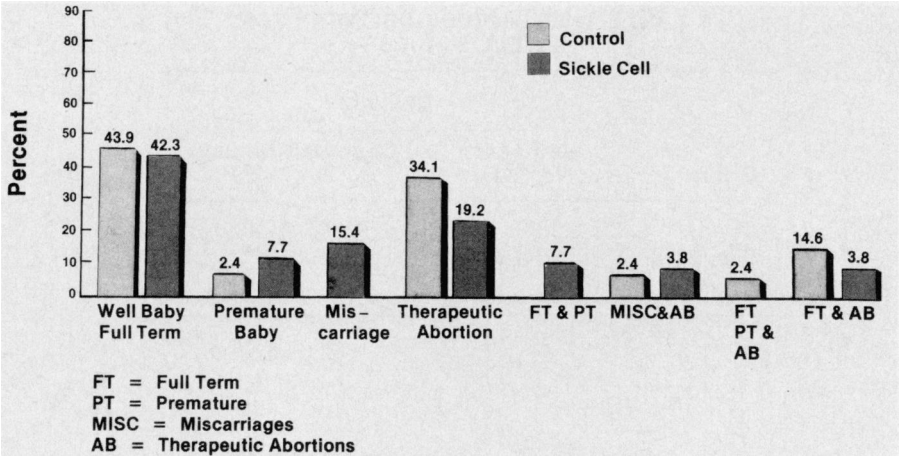

Figure 2. Pregnancy outcomes

Pregnancy

Both groups reported similar rates of conception (approximately 60 percent). The outcomes of the pregnancies were markedly different in that the sickle cell group experienced more miscarriages and premature births when compared with the controls ($P < .05$) (Figure 2).

There was a significant difference in the methods of delivery. Among the sickle cell group 46 percent reported cesarean sections as opposed to 18 percent in the controls. Therefore, a sickle cell mother had only a 54 percent chance of a normal vaginal delivery compared to an 82 percent chance for a mother in the controls (Table 1).

Abortion and Miscarriage

The mean age for an abortion or miscarriage was 20.8 years in the controls and 22.7 years in the sickle cell group. Both groups varied in age at the time of the abortion or miscarriage with ranges of 15 to 31 years and 15 to 43 years, respectively, in the controls and sickle cell patients. The mean length of gestation of the aborted fetuses was 2.4 months in the controls and 2.9 months in the study group. Gestational ages at the time of the abortion or miscarriage were 1 to 4 months in the controls and 2 to 5 months in the sickle cell group.

Children

The mean number of living children was 1.8 and 1.6 in the control and sickle cell groups, respectively (ranges of 1 to 6 in the control group and 1 to 5 in the sickle cell group). The age of the youngest child spanned 2 to 16 years (mean, 5.7 years) in the control group and 1 to 16 years with a mean of 9.9 years in the sickle cell group. The age of the oldest child in the sickle cell group ranged from 1 to 25 years (mean, 9.9) and in the control group, from 2 to 19 years.

Marital Status

Sixteen percent of the sickle cell patients and 18 percent of the controls were married; the remainder were divorced, separated, or single. Single status was similar in both groups (75 percent for the study group and 77 percent for controls).

TABLE 1. METHOD OF DELIVERY FOR WELL BABIES

Group	Delivery	
	Vaginal No. (%)	Cesarean Section No. (%)
Control	23(82.1)	5(17.9)
Sickle Cell	7(53.8)	6(46.2)

$\chi^2 = 2.89$, $P < .12$
$\Phi = .30$

DISCUSSION

Women with sickle cell disease in this study reported less sexual activity than the controls. In addition, members of the study group had been older at the time of their first sexual encounter. These patterns were also observed by Alleyne et al.[4] It has been proposed that delays in sexual maturation, the effects of the chronic disease on body habitus and function, and the psychological impact of the disease process on the individual may limit the exposure of these females to the opposite sex.[9] In this study, women with sickle cell disease reported less use of contraception. This could be directly related to the lower frequency of sexual activity. Whether there could be a psychological factor as well (eg, a need to prove their ability to reproduce) is a matter of speculation only; no data support such a conclusion. The sickle cell group used a greater variety of contraceptive methods than the controls, with many using the diaphragm and foam. The intrauterine device was also used more frequently in the sickle cell group. These findings may be a direct result of medical advice given to the sickle cell patients, who are often dissuaded from using birth control pills.

Adadevoh et al[10] reported low use of family planning clinics in Nigeria by women with hemoglobinopathies HbSS and SC. Foster[11] examined the risk-benefit ratio of the commonly used contraceptive methods and evaluated their application in sickle cell disease. The contraceptive methods evaluated were (1) steroid contraception, (2) intrauterine devices, (3) barrier methods, (4) timing ("rhythm") methods, (5) sterilization, and (6) abortion. He concluded that barrier methods combined with timing methods may offer the best balance. The increased incidence of thromboembolic phenomenon caused by combination oral contraceptives, in addition to the already increased incidence of thromboembolism in sickle cell disease, has led to the use of progestin-only contraceptives such as the mini-pill in patients with sickle cell disease.[10] Nevertheless, these estrogen-free contraceptives have not been exempted as causes of or contributors to the thromboembolic phenomenon. Of interest, however, is the report by Adadevoh and colleagues that progestogen preparations may have an antisickling effect.[10,12] In the present study sickle cell patients frequently used the intrauterine device despite the known hazards of complications due to pelvic infections and excessive bleeding. Perhaps factors such as age and maturity dictate the method of contraception in these patients as in the general population. The level of responsibility that an individual demonstrates must also be considered. The authors believe, therefore, that any recommendations for contraception in sickle cell patients must be individualized based on these criteria as well as those cited by Foster.[11]

The rate of conception was similar among the controls and the sickle cell group in the study. If fertility is defined as the ability to conceive, the

results of this study would not confirm reports of subfertility in sickle cell women.[4,5] The difference could be directly related to definitions used in each study; for instance, if fertility is regarded as the number of live births, results may vary. Even with this definition this study shows no difference in fertility. Dunne and Joseph[3] concluded in their study that fertility was normal in women with sickle hemoglobinopathies. Outcomes of pregnancy in terms of increased prematurity and miscarriage in the present study validate the numerous reports in which fetal wastage was excessive in sickle cell patients.[1,2,4] The specific cause for this finding needs further clarification. Factors to be considered are chronic anemia, status of maternal nutrition, and greater susceptibility to infection. Only the "well babies" were compared in this evaluation of the method of delivery, accounting for the reduced sample size. The greater number of cesarean sections was related to increased obstetrical complications. These included chronic anemia and such hip abnormalities as avascular necrosis of the femoral head, which caused pelvic abnormalities that aggravated the already contracted pelvis. The higher incidence of the surgical method of delivery may also reflect an increasingly prophylactic approach on the part of some obstetricians.

Finally, sickle cell traits were included as controls because of data by Pearson and Vaughan[8] showing a lack of influence of sickle cell trait on fertility and successful pregnancy. This finding was corroborated by Hendrickse et al.[2]

CONCLUSIONS

A study of 48 women with sickle cell disease revealed that these patients were less sexually active than the controls and used less contraception. In addition, the sickle cell patients were older at the time of their first sexual encounter. The ability to conceive was the same in both groups; however, the incidence of prematurity and miscarriage was greater in the sickle cell patients, who experienced more cesarean sections than the controls. The results of this study provide additional information that can be used in the prospective medical management and counseling of women with sickle cell disease.

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Literature Cited

1. Hendrickse JP, Watson-Williams EJ, Luzatto C, Ajabore LN. Pregnancy in homozygous sickle cell anemia. J Obstet Gynaecol Br Commonw 1972; 9(5):396-409.

2. Hendrickse JP, Watson-Williams EJ. The influence of hemoglobinopathies on reproduction. Am J Obstet Gynecol 1966; 94:739-748.

3. Dunne GD, Joseph RR. Fertility in hemoglobin S-S and hemoglobin S-C disease. Fertil Steril 1970; 21(8): 630-634.

4. Alleyne SI, Rauseo RD, Serjeant GR. Sexual development and fertility of Jamaican female patients with homozygous sickle cell disease. Arch Intern Med 1981; 141: 1295-1297.

5. Pearson HA, Vaughan EO. Lack of influence of sickle cell trait on fertility and successful pregnancy. Am J Obstet Gynecol 1969; 105(2):203-205.

6. Jimenez CT, Scott RB, Ferguson A, et al. Studies in sickle cell anemia: XXVI. The effect of homozygous sickle cell disease on the onset of menarche, pregnancy, fertility, pubescent changes and body growth in Negro subjects. Am J Dis Child 1966; 111:497-504.

7. Liu YK. Folate deficiency in children with sickle cell anemia. Am J Dis Child 1974; 127:389-393.

8. Olambiwonnu NO, Penny R, Frasier SD. Sexual maturation in subjects with sickle cell anemia: Studies of serum gonadotrophin concentration, height, weight, and skeletal age. J Pediatr 1975; 87:459-464.

9. Alleyne SI, Wint F, Serjeant GR. Psychological aspects of sickle cell disease. Health Soc Work 1976; 1:105-119.

10. Adadevoh BK, Dada OA. Contraception and haemoglobinopathies in Ibadan, Nigeria. An evaluation on the effect on anaemia. Trop Geogr Med 1977; 29:77-81.

11. Foster, HW. Contraceptives in sickle cell disease. South Med J 1981; 74(5):543-545.

12. Morrison JC. Sickle cell hemoglobinopathies and reproduction: International aspects of sickle cell disease. Proceedings of the First International Conference on Sickle Cell Disease: A World Health Problem. Washington, DC, 1976, pp 116-119.