Original Article

Sexuality and sickle cell anemia

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Background: *Sickle cell disease, the most common hereditary blood disease in the world, is the result of an atypical hemoglobin called S (Hb S) which, when homozygous (Hb SS) is the cause of sickle cell anemia. Changes of puberty, correlated with a delayed growth spurt, begin late in both male and female sickle cell anemia individuals with repercussions on sexuality and reproduction. The objectives of this exploratory and descriptive study were to characterize the development of sexuality in adults with sickle cell anemia by investigating the patient's perception of their sex life, as well as the information they had and needed on this subject.*

Methods: *Twenty male and female sickle cell anemia patients treated at the Hemocentro Regional de Uberaba (UFTM) with ages between 19 and 47 years old were enrolled. A socioeconomic questionnaire and a semi-structured interview on sexuality, reproduction and genetic counseling were applied.*

Results: *This study shows that the sickle cell anemia patients lacked information on sexuality especially about the risks of pregnancy and the possible inheritance of the disease by their children. Moreover, the sexual life of the patients was impaired due to pain as well as discrimination and negative feelings experienced in close relationships.*

Conclusion: *The health care of sickle cell anemia patients should take into account not only the clinical aspects of the disease, but also psychosocial aspects by providing counseling on sexuality, reproduction and genetics, in order to give this population the possibility of a better quality of life.*

Keywords: *Anemia, sickle cell/psychology; Sexuality; Quality of life; Humans; Adult*

Introduction

The clinical manifestations of sickle cell anemia (SCA), a chronic disease, occur within the first year after birth and extend throughout life with great variability of symptoms(1). The most frequent clinical problem is painful vaso-occlusive events(2); pain is a constant in the lives of adults(3).

Sexual development (involving physical and physiological aspects) in the general population accompanies bone age. In SCA patients, this development is delayed as they have delayed bone maturation due to the clinical manifestations of the disease. The anthropometric abnormalities of both male and female adult SCA patients have been well known for a long time. For example, menarche in SCA women is two to three years later than in non-SCA individuals with weight being a predictive factor(4).

Sexuality goes beyond the physiological aspects and has a direct relationship with the social environment in which one lives, with the emotions and feelings that relations produce permeating every moment of life(5). Sexuality is a historic and symbolic process that expresses the subject's identity and not only fulfills the function of social reproduction of the species but also involves the pursuit of personal pleasure(6). Sexuality is an essential characteristic of the human being, present in all stages of life(7) despite its assertion during adolescence. In the broad setting where sexuality intersects with SCA, there are other factors to consider such as the clinical emergency of priapism(8), the risk of pregnancy in women with SCA(9) and genetic counseling(10).

It is also important to remember that delayed pubertal development is a major source of stress for SCA patients. When puberty does not happen near the age of their peers, patients feel different and isolate themselves, at a time of life when social acceptance is of great importance. Feelings of inferiority and immature behavior have also been reported in respect to pubertal delay(11). Advances in the medical knowledge of SCA over the last ten years have increased the life expectancy of these patients(10). Therefore, studies on aspects that contribute to quality of life are needed, taking into account the concept of complete care as recommended by the World Health Organization (WHO)(12).

Thus, the aim of this study was to characterize the development of sexuality of SCA patients by investigating the representation (perception) they have about their sexual life and by identifying the information they have about sexuality, contraception, pregnancy and the genetics of SCA and their further information needs.

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The authors declare no competing financial interest

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Methods

This descriptive exploratory study, carried out in 2011, is the product of the residency program in Integrated Multidisciplinary Health, in particular adult psychology, a partnership between the Universidade Federal do Triângulo Mineiro (UFTM) and the Hemocentro Regional de Uberaba, Minas Gerais (Fundação Hemominas). This research was approved by the Ethics Committees of UFTM (# 1814) and Hemominas (# 309).

Twenty male and female patients diagnosed with SCA (hemoglobin SS) and aged between 19 and 47 years were recruited according to their medical records independent of racial background. Patients were contacted on the day of their medical appointments in the Uberaba Regional Blood Center. The nature of the study was explained and the consent forms were read to all prospective participants. After giving their consent, a sociodemographic and economic questionnaire was applied. The results were analyzed according to the Key Correction Classification of Socioeconomic Classes in Brazil which categorizes individuals into A, B, C, D, E and F socioeconomic classes(13). Subsequently, a semi-structured interview was conducted in order to characterize the sex life of the SCA patients, their representation of sexuality, as well as to describe the information they had or would like to have on issues related to their full development and the associated difficulties.

The interview was designed based on the literature found on the subject, taking into account the flexibility of a semi-structured instrument. This type of instrument was chosen because it provides a richer understanding of the population of interest. The semi-structured interview was designed with both closed and open questions, in which the informant was able to discuss the proposed topic(14).

The interviews were analyzed qualitatively using content analysis of Bardin(15). This analysis consists in transcribing the interviews and gathering content for themes relevant to the study. Similar reports of patients were grouped, but contradictions and ambiguities were also identified. Importantly, the analysis took place concurrently with the research. This is a process controlled by the researcher who has autonomy (following methodological criteria) to stop the interviews when it is noted that the interview will provide no new information (saturation criterion).

Therefore, the number of patients interviewed (n), determined by the data analysis method, must allow the researcher to understand the subject of the study well(16). This type of study defines the number of interviewees with progressive inclusion; inclusion is interrupted by the saturation criterion, that is, when the concepts, explanations and meanings attributed by interviewees start having a regular presentation(17).

Results

Sociodemographic data of the 20 patients are shown in Table 1. The majority of patients were women (75%), aged predominantly between 19 and 29 years (50%). Most were social class C (60%) and 45% were performing regular remunerated activities.

Concerning the sex life of the patients, 85% had already had intimate relationships. The first sexual intercourse happened between 10 and 36 years of age with most between 15 and 20 years (52.9%).

Table 1 - Sociodemographic data of 20 patients with sickle cell anemia

Variable	n	%
Gender		
Male	5	25
Female	15	75
Age group		
19-29 years	10	50
29-39 years	7	35
39-49 years	3	15
Social class*		
A	0	0
B	4	20
C	12	60
D	4	20
Employment		
Yes	9	45
No	8	40
Retired	3	15

*Key Correction Classification of Socioeconomic Classes in Brazil(13)

Eleven (55%) respondents considered that they had an active sex life with the frequency of sexual activity ranging from one to three times per week. Moreover, 82.3% of the patients rated their sexual activity as satisfactory; it was classified as excellent by 47.0% and as good by 29.4%. The majority of subjects (60%) had a partner in the previous year. Out of the 20 patients, 70% said they had had the desire to engage in sexual intercourse; 76.4% of those who had had intimate relationships said that they had enjoyed the experience. Most (65%) did not masturbate.

Thirteen (65.0%) of the 20 interviewees reported having impaired sexuality due to SCA or the associated clinical complications. In particular 12 (92.3%) of these 13 patients reported that the impairment was due to painful crises; the other patient reported fear of priapism (Table 2).

Eighteen (90%) patients had had affective relationships and all had informed the partner about their disease. Only two patients (11.1%) did not feel that they were supported by their partners. Moreover, they said that SCA influenced their sex lives (45%). Concerning their sexual characteristics, 55% of patients were compared to other people, as can be seen in Table 3. Some patients' statements illustrate the difficulties in interpersonal (affective and social) relationships.

In Table 4, 80% of men did not know about priapism. Two (20%) of five interviewed knew about the vasectomy contraceptive method. Only one reported having erectile dysfunction. In women, the age at menarche ranged from 12 to 20 years. Some were quite late, more often between 14 to 16 years. Menstrual cycles were reported by 53.3% women as regular. There were no reports of miscarriages.

Eight patients (40%) of 20 had had children. Most of them (75%) had only one child. No pregnancy was planned. The majority of the patients (60%) said they had knowledge about the risks of SCA for pregnancy, while two of them became aware only after becoming pregnant.

Table 2 - Sex lives of 20 patients with sickle cell anemia

Sex life	n	%
Sexual intercourse		
Yes	17	85.0
No	3	15.0
Age at first sexual intercourse*		
10-15 years	3	17.7
15-20 years	9	52.9
20-25 years	2	11.8
25-30 years	2	11.8
30-35 years	0	0
35-40 years	1	5.9
Sexually active		
Yes	11	55.0
No	9	45.0
Frequency of sexual activity (n°/week)		
0	9	45.0
1-3	8	40.0
> 3	3	15.0
Satisfaction with sex life*		
Yes	14	82.3
No	3	17.7
Evaluation of sex life*		
Great	8	47.0
Good	5	29.4
Regular	3	17.7
Bad	1	5.9
Loss of sexuality in sickle cell anemia or clinical complications		
Yes	13	65.0
No	7	35.0

*Patients who have had sexual intercourse (n = 17)

Table 3 - Perception of 20 patients with sickle cell anemia about their sexuality

Perception of sexuality	n	%
Their partner knew about sickle cell anemia		
Yes	18	100.0
No	0	0
Felt supported and cared for by their partner*		
Yes	16	88.9
No	2	11.1
Sickle cell anemia influenced their sex lives*		
Yes	9	45.0
No	11	55.0
Were compared to someone regarding their sexual characteristics*		
Yes	11	55.0
No	9	45.0

*Question were answered by those who had had a sexual partner

Half of the twenty patients had received genetic counseling and reported having knowledge on sexually transmitted diseases (STDs) (85%) and 65% of the patients had received general sex education.

Table 4 - Sexuality of 20 patients with sickle cell anemia according to gender

Variable			Variable		
Men	**n**	**%**	**Women**	**n**	**%**
Knew about priapism			**Age at menarche**		
Yes	1	20.0	12-14 years	5	33.3
No	4	80.0	14-16 years	6	40.0
Knew about vasectomy					
Yes	2	40.0	16-18 years	1	06.7
No	3	60.0	18-20 years	3	20.0
Episode of erectile dysfunction			**Regular menstrual cycles**		
Yes	1	20.0	Yes	8	53.3
No	4	80.0	No	7	47.7

Table 5 - Reproduction history of 20 patients with sickle cell anemia

Reprodution history	n	%
Children		
Yes	8	40.0
No	12	60.0
N° the children		
1	6	75.0
2	1	12.5
3	1	12.5
Planned pregnancy		
Yes	0	0
No	8	100.0

More than one source of information was mentioned by some of those interviewed. School was the main source of information on STDs and sexual education. Additionally, information about the prevention of STDs was obtained from family health teams. Specialized health teams linked to the second or tertiary services were the most commonly cited source for knowledge about the risks of pregnancy in SCA women and genetic counseling. However, the reported knowledge both on the risk of pregnancy in women with SCA and about genetic counseling was superficial, that is, without specifying causes or associations.

Table 6 - Sources of information according to the 20 patients of SCA

Sources of information	Risk of pregnancy	Genetic counseling	STDs	Sex education
Schools	0	0	9	11
Specialized health team	14	8	1	1
Family health team	0	0	4	1
Association	0	0	1	0
Media	0	0	1	0
Other	0	2	2	2

About contraception, 65% of patients had obtained information from specialized health teams. Use of contraceptives was reported by 50% with the most common method being the pill. On the other hand, 80% never used emergency contraception even though 60% knew about the method.

At the end of the interview, the patients were asked if they would like to have more information about sexuality. Six patients had doubts on the risk of pregnancy in women with SCA (2), genetic counseling (1), STDs (1), development and growth in SCA individuals (1) and about pleasure in sex (1).

Table 7 - Contraception use and knowledge of sickle cell anemia patients

Variable	n	%
Knowledge about contraception		
Yes	13	65.0
No	7	35.0
Source of information about contraception		
Schools	1	7.6
Specialized health team	7	53.4
Family health team (Brazilian health program)	1	7.6
Associations	0	0
Media	0	0
Other	4	30.4
Use of contraceptives		
Yes	10	50.0
No	10	50.0
Type of contraceptive*		
condom	3	30.0
Anti-conception pill	7	70.0
Knowledge of emergency contraception		
Yes	12	60.0
No	8	40.0
Use of emergency contraception		
Yes	4	20.0
No	16	80.0

*Answered by those who used contraceptive measures

Discussion

There was a predominance of females in the sample (75%) even though SCA is not more prevalent in women(16,18). This may be due to the fact that women tend to take better care of themselves and more frequently visit health services than men.

One publication(1) states that SCA is found in all social classes including class A. In this study though, most interviewees were in socioeconomic class C, which may be explained by the fact that this study was carried out in a government hospital and that SCA is more prevalent in people with lower socioeconomic conditions due to the history of SCA in Brazil with hemoglobin S arriving with the slave trade(17,19).

Analyzing the data about sexual relationships, patients with SCA had similar accounts to the general population. According to Brazilian Ministry of Health, 32.8% of Brazilians between 12 to 17 years old (most frequent response category) have already had sexual intercourse(18,20). In this sample, sexual relationships were frequent between 15 to 20 years old (52.9%). It is important to say that thirteen (65.0%) of the twenty patients reported impaired sexual relationships or activity due to SCA or its clinical complications, with pain being the main reason. Pain is also the most reported clinical complication in the literature(2,3).

Sexuality involves behavior and desires related to fulfillment, affection, pleasure, feelings, the exercise of freedom and health. It is a historical, cultural and social feature that is modified according to changes in social relationships(20). Thus, in the Brazilian society, sexuality is limited by taboo, myths, discrimination and power relationships(20). Adult patients with SCA feel that their chronic disease influences their sexuality. They compared themselves or were compared in respect to their sexual characteristics, which reflected on their relationship, feelings of discrimination, fear and insecurity. So, although as adults their sexual development is similar to individuals without the disease and they described having sexual relations similar to healthy people, SCA patients report discrimination and feelings of inferiority arising from their chronic disease and its consequences.

There was much ignorance (80%) about priapism which is of great concern as in the literature 28% to 38% of male patients have a history of priapism with the first episode occurring before the age of ten. Between 10% and 30% of these patients progressed to partial or complete erectile dysfunction, regardless of the type of therapy used and thus prevention is better than cure(8). Increased awareness of the signs of priapism and education about the need for early treatment are good ways to prevent long-term problems(21). However, priapism was not common in this population which may indicate good care.

Most participants (65%) reported having general knowledge about sex education; 85% had learnt about STDs and 65% about contraception. Although most indicated knowledge about the risks involved in pregnancy (60%) and about genetic counseling (50%) the information was very superficial, which was seen by the lack of family planning by the eight patients who had children. Therefore, there is a need for the healthcare system to extend sexual education in SCA.

Pregnancy in SCA often leads to complications in the health of both the mother and the child. For that reason, contraception linked to genetic counseling, enables adequate family planning and is an important factor in the health care of women with SCA(9).

Genetic counseling is an important component of the clinical management of patients with SCA; this involves significant medical, psychological, social, ethical and legal implications. The goals should be primarily educational and healthcare, that is, enable individuals or families to make consistent and psychologically balanced decisions about procreation(10).

A need to expand counseling for individuals with SCA is clear from the results by increasing accessibility in order to break with superficiality, extrapolating the "taboo" of sexuality and making it what it should be: a natural and healthy process.

Finally, it is worth noting that the embarrassment in answering questions related to sexuality was constant in interviews, showing that, despite a rapid development in sexual relationships, with greater acceptance and a broader discussion in society as a whole, there are remnants of judgments and social impositions that individuals stay silent on the subject of sexuality.

Conclusion

In addition to the clinical care of the illness, healthcare measures focused on the patient with SCA should aim to take into account the normal development of the individual providing consistency between personal needs and the needs related to the disease. One must stress the information on the social expectations of an ordinary person, such as is the case of sexuality. It requires not only information on self care and social representation, but also on reproductive care. These actions should guarantee the integrity of health as advocated by the WHO.

It is important to point out that public health policies do not focus on chronically sick people. And so the access of this population must be expanded to existing public policies. It is noteworthy that in Brazil, although public health policies have proposed a new care model, curative treatment still prevails for SCA with care and treatment being provided in the emergency room or at hospital admissions and preventive care being considered of little importance.

It seems that there is little acceptance or discussion on the topic of sexuality. One of the actions of a multidisciplinary team or the professional psychologist is to listen to the personal and social needs of the patient that go beyond the disease. Professional care, including the provision of information and prevention and intervention strategies, can be carried out for individual cases or in groups.

Finally, it appears that there are few Brazilian studies that analyze the quality of life of people with SCA suggesting that more research and studies on the chronicity of the disease and its repercussions are needed.

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