

MEN'S SEXUAL HEALTH

Priapism in Sickle Cell Disease: An Evaluation of the Knowledge of an at Risk Population in Jamaica



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ABSTRACT

Introduction: Ischemic priapism is characterized by painful erections that may lead to erectile dysfunction. Men with sickle cell disease (SCD) are particularly prone to this condition, however, the knowledge among this population with respect to possible complications is not well known.

Aim: The objective of this study was to evaluate the knowledge of males with SCD about priapism and its possible consequences.

Methods: A cross-sectional study was carried out among consecutive consenting males aged 12 years and older with SCD presenting to the Sickle Cell Unit between September 2018 and August 2019. All participants completed a questionnaire detailing knowledge on the definition of priapism, its association with SCD, consequences of untreated priapism and treatment strategies. The responses were used to generate a total priapism knowledge score for each of the participants.

Main Outcome Measures: Main outcomes included knowledge of the term priapism, its association with SCD as well as the total priapism knowledge score.

Results: 219 patients of mean age 29.8 ± 13 years completed the questionnaire. 38.4% of patients were familiar with the term priapism and of these 68.8% were aware of the association between SCD and priapism. There was a significant association between knowledge of association of priapism with SCD and increasing educational level ($P = .036$) and history of prior priapism episodes ($P = .02$). There was a significant association between knowledge of the term "priapism" and history of priapism ($P = .002$). The mean total priapism knowledge score among the participants was 5.16 out of a maximum score of 12, with 70.8% of participants being categorized as having "poor knowledge."

Conclusion: There is a general lack of knowledge among patients with SCD with respect to the term priapism. Education campaigns geared toward addressing the identified knowledge gaps may prove beneficial in increasing awareness among this population and potentially decrease the risk of complications. **Whyte N, Morrison-Blidgen B, Asnani M, Priapism in Sickle Cell Disease: An Evaluation of the Knowledge of an at Risk Population in Jamaica. Sex Med 2021;9:100339.**

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Key Words: Priapism; Sickle cell disease; Priapism knowledge; Priapism complications

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INTRODUCTION

Priapism is defined as a full or partial unwanted erection that lasts beyond 4 hours after sexual stimulation and orgasm or is unrelated to sexual intercourse.¹ The condition is broadly classified into two types: ischemic (low flow) and non-ischemic (high flow, arterial). Ischemic priapism is characterized by a persistent erection with rigidity of the corpora cavernosa with little or no cavernosal inflow and is also associated with pain. Patients with

sickle cell disease are prone to having episodes of ischemic priapism as well as a variant of the condition known as stuttering priapism, which describes a pattern of recurrence of episodes of short duration.¹ The lifetime probability of a man with sickle cell disease developing ischemic priapism ranges from 29 to 42%.² In a study of 162 Jamaican males with sickle cell anemia, a cumulative incidence of almost 60% of this population had experienced priapism by the age of 40 years old.³ Of these, many were only elicited after direct questioning partly because of lack of knowledge that priapism could be a complication of sickle cell disease.

The mechanism underlying ischemic priapism is that of an imbalance between vasoconstriction and vasorelaxation within the penis.¹ During episodes of ischemic priapism, blood becomes trapped within the corpora cavernosa causing tissue hypoxia, hypercarbia as well as acidosis. This situation lends itself to a painful compartment syndrome type picture and if same is not adequately treated it can lead to myonecrosis and eventually fibrosis. The end result is a potentially disfigured penis with erectile dysfunction that is refractory to treatment, save for a penile prosthesis which becomes a technically more difficult procedure due to scarring.⁴ A 2002 study by Adeyoju et al, noted that among patients who experienced priapism, 21% reported having subsequent erectile dysfunction.⁵ The risk of irreversible complications increases with increasing time to presentation. For these reasons, ischemic priapism is considered a medical emergency.⁶

Jamaica represents a population in which the spectrum of sickle cell disease is common with 10% of the population having the sickle cell trait.⁷ It is important for patients affected by the disease to be aware of their risk of priapism and be active in the management in an effort to prevent or decrease the effect of complications from the condition. In general, there is limited data on the knowledge of patients with sickle cell disease about priapism. Previous studies have been done in Nigerian, Senegalese and Togolese populations assessing knowledge of patients with sickle cell about priapism. These revealed that most were not knowledgeable about priapism and most were also unaware of the link between sickle cell disease and priapism.⁸⁻¹⁰

Currently, there are no studies looking specifically at the Jamaican population of patients with sickle cell disease with respect to their knowledge of priapism. The authors hypothesized that knowledge among sickle cell patients about priapism and its complications is poor. Given the fact that Jamaica represents a population in which there is a high burden of sickle cell disease we aimed to evaluate the knowledge of this at risk population on priapism and its possible consequences.

MATERIALS AND METHODS

A single-center cross-sectional study was carried out among consecutive consenting males aged 12 years and older with sickle cell disease who presented to the Sickle Cell Unit of the University of the West Indies, Kingston, Jamaica between September 2018 and August 2019. Ethical approval was obtained from the

University of the West Indies (UWI) Ethics Committee, Kingston, Jamaica. Exclusion criteria were patients less than 12 years of age, female patients and persons acutely ill. Persons who were already actively involved in more than one study were also excluded. The projected sample size was 235 participants (5% error; 95% confidence level). This was calculated based on the number of active male patients over 12 years in the Sickle Cell Unit's database (1268), and assuming 75% prevalence of "poor knowledge".

Consecutive eligible patients presenting to the Unit were informed of the study by a research assistant and invited to participate. Written informed consent was obtained from patients (and parent(s)/ guardians of children under age 16 years) who agreed to participate. The research assistant administered a one-time questionnaire to all participants, either before or after a routine ambulatory care visit. The questionnaire consisted of fifteen questions, the majority of which were multiple choice. These questions served to gather basic demographic data and also gathered data on the participants' knowledge of the term priapism, its association with sickle disease, consequences of untreated priapism and ideal treatment strategies. There were eight specific knowledge-based questions which were used to generate a knowledge score. Two of these eight questions had sub-sections. Each of the specific knowledge-based questions was allocated one point with the exception of the two questions with sub-sections in which each section within the question was given a score value of one point. This enabled the authors to create a knowledge score with a maximum possible knowledge score of 12. A score of less than 6.2 (based on mean + $\frac{1}{2}$ standard deviation cut-off priapism knowledge score) was considered as poor knowledge. The questionnaire was created by one of the authors and reviewed by co-authors and other experts in the field of urology and sickle cell disease. The questions were also reviewed and approved by the UWI ethics committee. To our knowledge, no previous standardized questionnaire existed to assess knowledge among patients with Sickle Cell Disease. The interview was conducted in a private area to maximize patient comfort and privacy and took approximately 15 minutes to complete.

Data were analyzed using IBM SPSS version 26. Responses were expressed as frequencies. Data was summarized as proportions. Analysis was done using the Chi-Square test for binary variables. A *P* value less than .05 was considered significant. Knowledge score was dichotomized as 'good' or 'poor' knowledge based on a cut-off of mean + $\frac{1}{2}$ SD knowledge score. Multivariate logistic regressions were performed to identify predictors of good knowledge for children and adults separately.

RESULTS

A total of 219 participants completed the questionnaire. Most of the patients had a self-reported sickle cell genotype of homozygous HbSS, 76.6% (*n* = 167), followed by HbSC 18.3% (*n* = 40). The remainder had HbSβ+, 2.8% (*n* = 6), another beta

Table 1. Patient demographics and genotype, priapism knowledge, and priapism history

Parameters	
Total number of respondents	219
Mean (SD) age, years	29.80 (13.325)
Genotype n (%)	
HbSS	167 (76.6)
HbSC	40 (18.3)
HbS β +	6 (2.8)
Other	2 (0.1)
Unknown	3 (1.4)
Educational level n (%)	
Primary	28 (12.8)
Secondary	128 (58.4)
Tertiary	63 (28.8)
Knowledge of term priapism n (%)	
Yes	84 (38.4)
No	135 (61.6)
History of priapism n (%)	
Major priapism n (%)	35 (16)
Stuttering priapism n (%)	78 (35.6)
Medical attention n (%)	
Time to medical attention n (%)	36 (40)
4 hours or less	13 (39.4)
>4 hours to 24 hours	10 (30.3)
>24 hours to 48 hours	5 (15.2)
>48 hours	5 (15.2)

chain variant, 0.9%, ($n = 2$), or did not know the type of sickle cell disease they had, 1.4% ($n = 3$). The mean age of the respondents was 29.8 years \pm 13.3 years (Table 1). Of these, 21.9% ($n = 48$) were children (ie, age <18 years). The majority of the sampled population, 58.4% ($n = 128$), had secondary level education while 28.8% ($n = 63$) and 12.8% ($n = 28$) had tertiary and primary level education respectively (Table 1).

Of the respondents, 61.6% ($n = 135$) did not have any knowledge of the term “priapism” (Table 1). After the term was explained, 68.8% ($n = 150$) thought that there may be a risk of developing priapism given the diagnosis of sickle cell disease; however, 22.5% ($n = 49$) were unsure and 8.7% ($n = 19$) thought there was no association between the two conditions.

Among the participants, 41.6% ($n = 91$) reported having a history of priapism; 16% ($n = 35$) had previously experienced a major priapism episode and 35.6% ($n = 78$) had experienced stuttering priapism. Among all patients who experienced priapism, 60% ($n = 54$) did not seek medical attention with episodes of priapism. Of the patients who did not seek medical attention with episodes of priapism, 77.8% ($n = 42$) had stuttering priapism. Among those who sought medical attention, 60.7% ($n = 20$) presented after 4 hours had elapsed with 30.2% ($n = 5$) presenting after 24 hours (Table 1).

It was found that 63.9% ($n = 140$) of the participants thought there may be a risk of irreversible complications associated with

priapism and a corresponding proportion, 63% ($n = 138$) thought this risk was time dependent. 54.1% ($n = 118$) of patients believed priapism could lead to erectile dysfunction however 45.9% ($n = 100$) did not identify erectile dysfunction as a complication of priapism (Figure 1).

It was found that there was a significant association between knowledge of association of sickle cell disease with priapism and increasing educational level ($P = .036$) and history of prior priapism episodes ($P = .020$). There was also a significant association between knowledge of the term priapism and a patient having a prior history of experiencing priapism ($P = .002$). Knowledge of the term priapism was significantly associated with increasing educational level ($P < .001$).

The mean total priapism knowledge score among the participants was 5.16 (Minimum 0; maximum 11) (Figure 2). On categorizing knowledge score into good and poor knowledge (based on cut-off of mean + $\frac{1}{2}$ SD), 70.8% of the participants had poor knowledge and 29.2% had good knowledge. On bivariate analyses, there was a significant positive correlation between age and total priapism knowledge score, $r(215) = .14$, $P = .044$. Similarly, there was a positive correlation between total priapism knowledge score and level of education, $r(217) = .16$, $P = .016$ with highest knowledge scores being seen among those with tertiary level education. Males with a history of priapism had a higher total priapism knowledge score than those who did not and this was statistically significant ($P = .022$).

On multivariate analyses, conducted separately for adults and children, it was found that for adult males, not having a history of priapism lowered the odds of good knowledge almost by half (OR: 0.45; P value: .020). In children, the only significant relationship was seen with genotype where those boys with genotype SC disease were more likely to have good knowledge than boys with SS genotype (OR: 16.1; P value: .049).

DISCUSSION

This is the first study to assess knowledge among Jamaican males with sickle cell disease about priapism. The number of participants in this study ($n = 219$) was larger in comparison to most of the other similar studies done in other parts of the world. There was one study among a Senegalese population which had 221 participants overall, however only 108 of these patients had sickle cell disease and the remainder comprised a control group of 113 healthy participants.⁹

The prevalence of priapism among our participants of 41.6% parallels the previously quoted figure of 42% among Jamaican males with sickle cell disease.² This prevalence rate is also similar to the 39.1% prevalence reported in a Nigerian population but was much higher than the 26.3% prevalence of priapism reported in a Togolese population.^{8,10}

Previous studies done in different populations assessing the knowledge of patients with sickle cell disease revealed generally

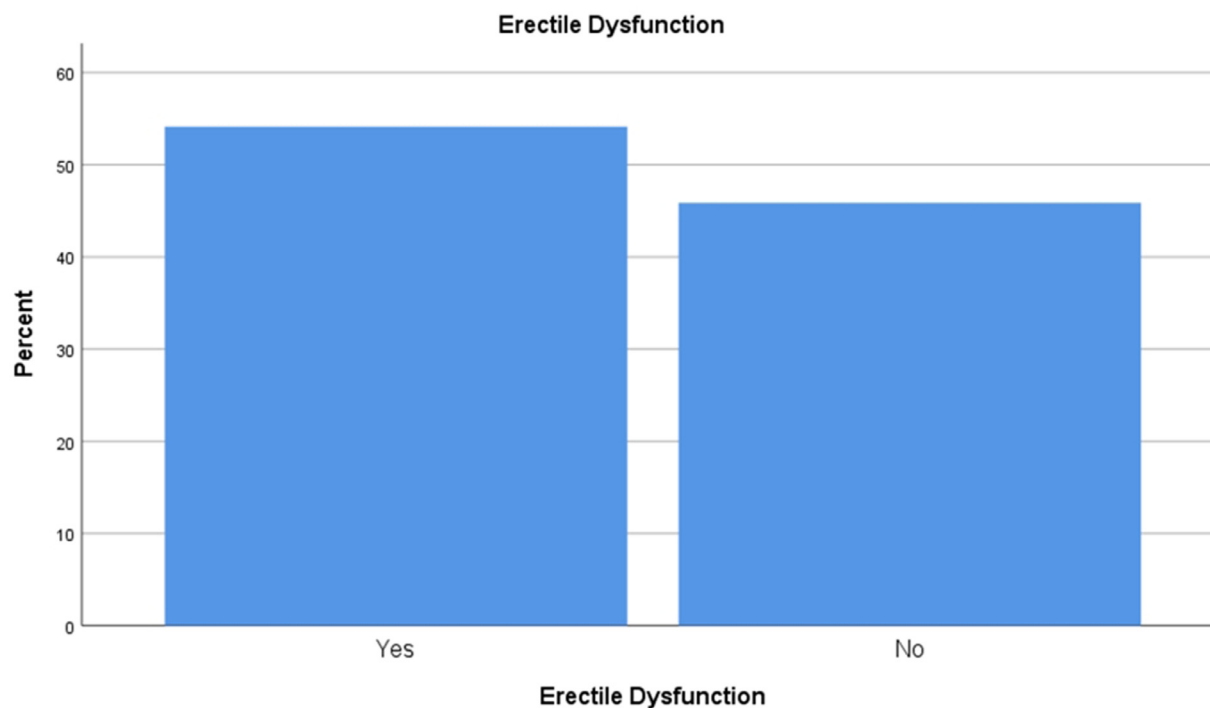


Figure 1. Bar chart showing knowledge among participants of erectile dysfunction as a complication of priapism.

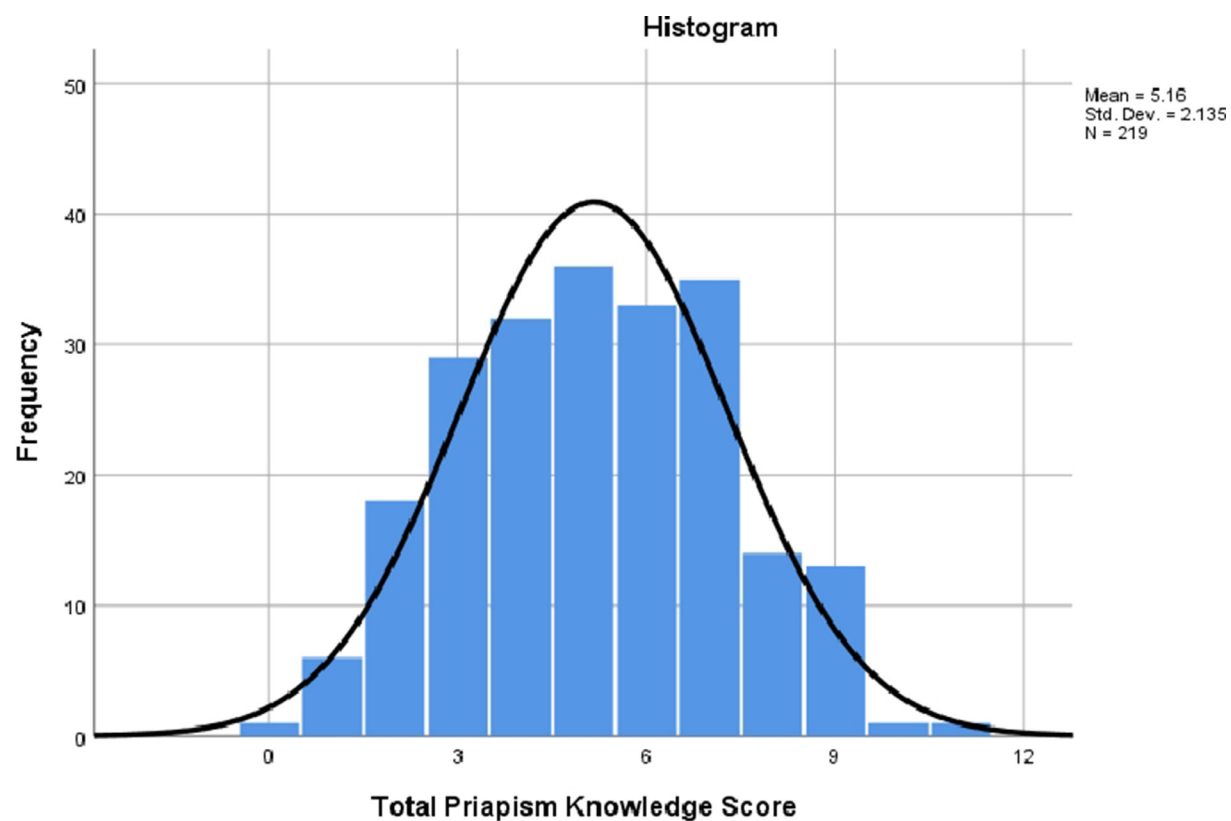


Figure 2. Histogram showing total priapism knowledge scores among participants.

that most did not know about priapism and most were also unaware of the link between sickle cell disease and priapism. In a cross-sectional study consisting of 114 Nigerian patients with homozygous sickle cell disease, only 21% were knowledgeable about priapism and 67.5% of them were not aware of the risk of priapism in sickle cell disease.⁸ Similar findings were seen in a study of patients in Senegal wherein of 108 patients with sickle cell anemia, 10.2% knew about priapism and 75% of that population did not know of an association between their disease and priapism.⁹ Another study from a Togolese population revealed that only 15.8% of the homozygous population without a prior history of priapism had ever heard of the condition.¹⁰

Our findings also showed that there was a general lack of knowledge among patients with sickle cell disease with respect to priapism, with the majority of the participants, 61.5%, being unfamiliar with the term priapism. However, this lack of knowledge appeared to be to a slightly lesser degree than in similar studies outlined above. These findings may be due to the fact that the sampled population were patients who were seen in a dedicated sickle cell unit. Patients who are routinely followed up at such a facility may be more likely to have knowledge of complications of sickle cell disease including priapism as patient education may be routinely done by health care providers. It should be noted that although knowledge of the term priapism was slightly higher than in other studies, the mean total knowledge score calculated among the participants generally reflected poor knowledge of the condition. The extent of poor knowledge in this respect could not be directly compared to other populations as previously, a standardized knowledge score for priapism did not exist. However, one Nigerian study reported categorizing its participants into good and poor knowledge and found that among its 95 participants, 55.8% had good knowledge of priapism which differs from our findings.¹¹

Although knowledge of priapism and its association with sickle cell disease was found to be poor among the sampled population, it was noteworthy to find that people of a higher educational level were more likely to be aware of priapism and its aforementioned link with their disease. There was also a significant positive correlation between education and the total priapism knowledge score, with higher scores being found among participants with tertiary level education. These findings suggest that level education is an important factor influencing knowledge of priapism among patients with sickle cell disease.

Despite the finding that the subset of patients who previously experienced priapism were among the ones to be more likely to have knowledge of the term priapism as well as its association with sickle cell disease, it did not seem to be associated with that subset having a good grasp of overall knowledge of the condition with respect to its complications nor did it translate into better health seeking behaviors in relation to treatment of the condition. This was evidenced by the fact that the majority of patients who experienced priapism did not seek medical attention for same. It should however be noted that a significant proportion of

those who did not seek medical attention had stuttering episodes and that such episodes may abate before the need for emergency room visits. Further details surrounding these stuttering episodes were not explored in the study and may have been useful in identifying those who might have benefited from emergency medical care. Among those who sought medical attention, most presented after four hours and just under one third presented after an extremely delayed interval of more than 24 hours. This tendency towards a prolonged interval between onset and seeking medical care was also seen in unpublished Jamaican data wherein the mean time to presentation to the hospital was found to be 20.6 hours (Morrison B, Rhudd A. Major priapism at the university hospital: 2000-2010. Unpublished data).

This delay in presentation to the hospital with episodes of priapism may suggest that patients may not be aware of the time dependent nature of risk of complications associated with this condition. However, when participants were asked specifically about this, our results showed that the majority of participants thought there was a time dependent risk of irreversible complications associated with priapism. The reason for this discrepancy between the patients' opinions on this and what was actually practiced was not identified by the study. It may be that there are other factors at play informing their practice, for example embarrassment but this cannot be conclusively stated since it was not explored. Perhaps this is an area that can be investigated in future studies.

It is worth mentioning that although the majority of patients thought that there was a risk of irreversible complications associated with priapism, a notable proportion, 45.9%, were not specifically aware of erectile dysfunction being one such complication. One may postulate that if patients knew the specific complications of untreated priapism, including erectile dysfunction, it may lead them to make more informed decisions in regard to seeking health care for this condition.

There are some limitations in this study that could be addressed in future research. One such limitation is the fact that the questionnaire which was administered to the participants was not pretested with the sample population before widespread use. This would have aided in identifying, for example, questions which may have been ambiguous and could therefore affect the responses. This could potentially affect the validity and reliability of the measurement tool and its ability to sufficiently measure the outcomes of interest. Another limitation is the sample size. Although it was larger than most other similar studies, it was less than what was projected to sufficiently power the study.

CONCLUSION

In conclusion, our study found that there was general lack of knowledge among the sampled Jamaican males with sickle cell disease with respect to priapism. Given that Jamaica represents a population in which sickle cell disease is so prevalent, it may prove beneficial to formally evaluate how patients with sickle cell

are currently being educated about complications associated with the disease, specifically priapism. Following from this, a targeted public education campaign can be designed in an effort to increase effective education among this vulnerable population with respect to priapism and ways to mitigate against long term sequelae of the condition.

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STATEMENT OF AUTHORSHIP

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