

Priapism is Linked to Low Quality of Life (QoL) in Nigerian Cohorts with Sickle Cell Anaemia

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Abstract

Background: Priapism, a notorious cause of painful and psychological stress in males with Sickle Cell Disease (SCD), has significant potential to negatively impact the overall Quality of Life (QoL). This study aimed to appraise QoL among adults with SCA-related priapism.

Methods: An unmatched case-control study involving 100 adult males with SCA (aged 16–45 years) was conducted at two tertiary hospitals in Kano, North-Western Nigeria, from January 2023 to June 2024. Clinical data were obtained and QoL was determined using the RAND 36-Item Health Survey (Version 1.0). Statistical analysis was performed using SPSS v21.0.

Results: The socio-demographics of the two participant groups were statistically similar ($p > 0.05$). Participants with priapism reported significantly lower scores across all domains: physical functioning ($p = 0.02$), physical health ($p = 0.04$), emotional health ($p = 0.01$), energy ($p = 0.04$), emotional well-being ($p = 0.04$), social functioning ($p = 0.04$), pain ($p = 0.03$), and general health ($p = 0.03$). Among the SCA patients with priapism, 13 (26%), 17 (34%), and 20 (40%) had mild, moderate, and severe clinical forms of priapism, respectively, and those with severe priapism reported lower scores in most domains of the SF-36 ($p < 0.05$).

Conclusion: Sickle cell anaemia-related priapism is associated with low Health-Related Quality of Life (HRQoL) across all domains of the RAND 36-Item Health Survey, and the QoL worsens with increasing severity of priapism in this cohort of Nigerian adults with SCA.

Keywords: QoL; SCA; priapism; clinical severity.

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Introduction

Priapism refers to a recurrent disorder of prolonged, undesired, painful penile erection with or without sexual stimulation.^{1,2} It is estimated to occur in 40% of males with SCD during their lifetime, making it a common morbidity in this population.³⁻⁵ Regrettably, despite various coping strategies employed, afflicted persons often continue to experience shame, anxiety, depression, and declined sexual function.³⁻⁵

This unfavourable experience resulting from priapism can negatively affect the overall health of men with SCD, encompassing their physical, mental, and social well-being, with attendant impact on their QoL. Several studies have previously evaluated QoL in SCD patients using various screening tools, either without any predictor or using selected clinical determinants such as blood transfusion, vaso-occlusive crisis, and foetal haemoglobin, among others.⁶⁻⁹ Lower QoL across many domains of the HRQoL questionnaire was reported among Nigerian patients with SCD compared to their counterparts without the disease.^{10,11} With the exception of priapism, most of the clinical parameters contributing to low QoL among Nigerians with SCD have been thoroughly evaluated by various studies.¹²⁻¹⁴ The Priapism in Nigeria (PIN) study, currently the most comprehensive published work on the clinical epidemiology of priapism and sexual dysfunction in Nigerian men with SCD, utilised the Priapism and International Index of Erectile Function (IIEF) questionnaires.^{4,5} Therefore, because the PIN study focused primarily on priapism itself and its

consequences on sexual health, it did not provide a holistic assessment of the effect of priapism on the overall Health-Related Quality of Life (HRQoL) in men with SCD in Nigeria.

The Medical Outcome Study (MOS) 36-Item Short Form Survey Instrument (SF-36) measures both physical and mental components of HRQoL, where a lower score denotes a worse limitation imposed on the individual's life by the condition under study in a particular SF-36 domain.¹⁵ The utility of the SF-36 has been validated in the assessment of HRQoL in individuals with SCD, and several studies worldwide have used it to evaluate the restriction imposed by SCD and its complications on the QoL of affected persons.^{16–19}

In an attempt to fill this knowledge gap regarding the role of priapism in the overall health status of adult men with SCD in Nigeria, this study used the RAND 36-Item Health Survey (Version 1.0) and its scoring tool to investigate the hypothesis that SCA-related priapism is a cause of low quality of life in Nigerian men.

Materials and Methods

An unmatched case-control study with a 1:1 case-to-control ratio, involving 100 adult males with SCA, was conducted at the Haematology Departments of Aminu Kano Teaching Hospital and Murtala Mohammed Specialist Hospital, Kano, North-Western Nigeria, from January to June 2024. Individuals diagnosed with SCA using haemoglobin electrophoresis on a cellulose acetate membrane in an alkaline medium or High-Performance Liquid Chromatography (HPLC), and who were in the steady state, were consecutively recruited as follows:

Cases: those who had at least three (3) episodes of priapism in the past year.

Controls: those who had no history of a priapism episode in the past year.

Bio-demographic and clinical data of the participants were obtained using a structured interviewer-administered questionnaire, and clinical notes from the patient folder were used to cross-check for any disparity of information. Quality of life was determined using the RAND 36-Item Medical Outcome Study (MOS) Short Form Survey Instrument (SF-36) (RAND® Corporation, California, USA) and processed according to its scoring protocol.

A composite scoring system combines data from multiple variables or items into a single score, providing a concise representation of a complex concept or performance.²⁰ We devised a composite scoring system, code-named the FDF Composite Score for clinical severity of priapism. The name FDF denotes the three parameters used to generate the score, as depicted in Table 1: Frequency of priapism in the past year, Duration of the longest priapic episode in the past year, and Form of intervention employed for the longest priapic episode. Frequency of priapism in the preceding year of <3, 3–6, and >6 episodes was assigned a score of 1, 2, and 3, respectively. Duration of the longest priapic episode in the preceding year of <4 hours, 4–6 hours, and >6 hours was assigned a score of 1, 2, and 3, respectively, while form of intervention employed for the longest priapic episode—medication, aspiration, and surgery—was assigned a score of 1, 2, and 3, respectively.

Data were analysed using the Statistical Package for the Social Sciences (SPSS) version 21.0 (IBM, Chicago, USA). The Kolmogorov–Smirnov test was used to assess the normality of data distribution. An independent t-test and ANOVA were employed, as appropriate, for the comparison of mean scores, while chi-square (χ^2) analysis was used to compare categorical variables.

The research proposal and its conduct were approved and supervised by the Research Ethics Committee (REC) of Aminu Kano Teaching Hospital (AKTH), Kano (Reference number :

Table 1. FDF Composite Score for clinical severity of priapism.

Element of composite score	Mild	Moderate	Severe
Frequency of priapism in the preceding year	<3 (1)	3–6 (2)	>6 (3)
Duration of longest priapic episode in the preceding year (hours)	<4 (1)	4–6 (2)	>6 (3)
Form of intervention employed for longest priapic episode	Medication (1)	Aspiration (2)	Surgery (3)

FDF Composite Score (Frequency, Duration, Form of intervention). The individual score for each patient is derived from the average of the three composite-score elements; the highest obtainable average score for the clinical categories of mild, moderate, and severe is 1, 2, and 3, respectively. Scores of 0–1.00, 1.01–2.00, and above 2.00 were categorised as mild, moderate, and severe forms of clinical priapism, respectively.

NHREC/28/01/2020/AKTH/EC/2906 via AKTH/MAC/SUB/12A/P-3/VI/3006). Informed consent was obtained from each participant, and all procedures were performed in compliance with the principles of the Declaration of Helsinki for experimentation involving human subjects.

Results

The socio-demographics of the two groups of study participants (cases and controls) and the clinical severity of priapism amongst cases are depicted in Table 2, wherein no statistically significant difference existed in the socio-demographic characteristics of the participants, and 13 (26%), 17 (34%), and 20 (40%) had

mild, moderate, and severe priapism, respectively.

The participants with priapism had lower scores than their counterparts without priapism across most domains of the MOS: physical functioning, 55.64 ± 28.17 versus 68.72 ± 17.62 ($p = 0.02$); emotional health, 48.08 ± 50.13 versus 70.10 ± 36.51 ($p = 0.01$); energy, 50.64 ± 6.20 versus 55.64 ± 4.89 ($p = 0.04$); and social functioning, 52.95 ± 23.86 versus 58.65 ± 16.16 ($p = 0.04$), among others, as depicted in Table 3.

Amongst participants with priapism, clinical severity of priapism is also related to low QoL scores, as those

Table 2. Socio-demographic characteristics of the participants and clinical severity of priapism amongst cases.

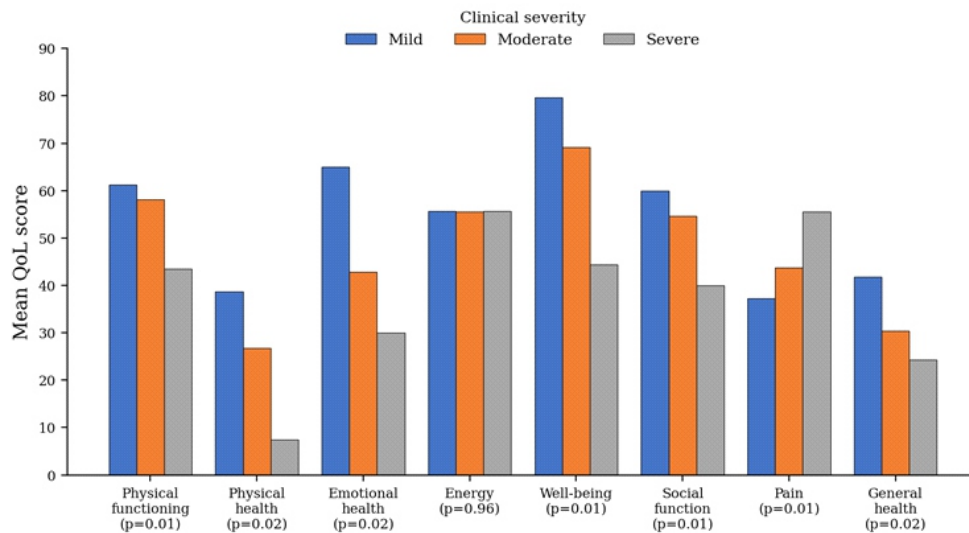
Socio-demographics	Cases (SCA with priapism) N = 50	Control (SCA without priapism) N = 50	P-value
¥Age (years)	23.64 ± 4.62	21.40 ± 4.44	0.17
¥Monthly income (₦)	208,506.08 ± 92,432.50	199,987.12 ± 87,385.11	0.68
≠Ethnic group			0.25
Hausa	42	39	
Igbo	3	3	
Yoruba	4	5	
Others	1	3	
≠Marital status			0.09
Single	48	46	
Married	2	4	
≠Education			0.15
Tertiary	2	0	
Secondary	28	27	
Primary	19	23	
Non-formal	1	0	
≠Occupation			0.13
Student	43	44	
Civil servant	4	5	
Business	0	1	
Unemployed	3	0	
#Clinical severity of priapism amongst cases			
Mild	13 (26%)		
Moderate	17 (34%)		
Severe	20 (40%)		

¥ = Student t-test; ≠ = chi-square analysis; # = composite-score system.

Table 3. Quality of Life (QoL) of the participants.

QoL domain	Cases (with priapism) Mean $\pm$ SD	Control (without priapism) Mean $\pm$ SD	P-value
Physical functioning	55.64 $\pm$ 28.17	68.72 $\pm$ 17.62	0.02
Physical health	26.41 $\pm$ 35.89	30.13 $\pm$ 29.34	0.04
Emotional health	48.08 $\pm$ 50.13	70.10 $\pm$ 36.51	0.01
Energy	50.64 $\pm$ 6.20	55.64 $\pm$ 4.89	0.04
Emotional well-being	63.79 $\pm$ 15.85	61.03 $\pm$ 9.73	0.04
Social functioning	52.95 $\pm$ 23.86	58.65 $\pm$ 16.16	0.04
Pain	45.19 $\pm$ 24.30	51.35 $\pm$ 15.95	0.03
General health	31.21 $\pm$ 11.29	38.82 $\pm$ 10.11	0.03

SD = standard deviation.

Figure 1. Comparison between Quality of Life (QoL) scores and clinical severity of priapism amongst participants with priapism (ANOVA with post-hoc analysis).

with severe priapism had significantly lower scores which persisted after post-hoc analysis across all domains of the MOS except energy ($p = 0.96$), as depicted in Figure 1.

Discussion

Health-Related Quality of Life (HRQoL), though often neglected, deserves serious attention in the management of SCD. This is particularly true given the continuous advances in SCD care, which have led to a life expectancy comparable to that of persons without the disease.^{5,7,13} Assessing HRQoL allows us to understand the burden of SCD and its complications, such as priapism, on the patients' overall well-being from their own perspective, rather than relying on healthcare personnel's perceptions. This approach will ensure that the prolonged life is free from the limitations imposed by SCD and is ultimately judged

by the affected persons as worth living.

This study reported poor HRQoL among male patients with SCA-related priapism, demonstrated by low scores across all domains of the MOS Short Form Survey. Importantly, QoL worsened with increasing severity of priapism. Our findings suggest that priapism negatively impacts the physical, social, and psychological health of adult males with Sickle Cell Disease (SCD).

While several studies have reported various clinical and epidemiological characteristics that contribute to poor QoL in SCD patients, to the best of our knowledge, no previous study specifically evaluated the burden of priapism on the overall HRQoL of males with SCA in Nigeria.

This gap highlights the persistent disregard of priapism as a serious complication of SCD, despite its potential to affect self-image. This oversight may be among the reasons why nearly 50% of patients with SCD-related priapism have never sought medical attention, as previously reported.⁵ It is noteworthy that a study from Lagos, Nigeria, which reported that priapism affected only the physical functioning domain of the SF-36, was primarily evaluating HRQoL among adult SCD patients generally and was not specific to priapism.¹³ As such, that finding might represent a mere casual association, potentially influenced by other SCD complications like bone pain crisis and acute chest syndrome, which received more prominence in the study.

Our findings of gross affectation across all domains of the SF-36 by priapism are not unexpected. The typical persistent, painful penile erection in priapism can readily interfere with the physical functioning, physical and general health, pain, and energy domains. Furthermore, the reported sadness, embarrassment, and fear in men with SCD-related priapism can directly interfere with the energy, emotional well-being, and social functioning domains of the SF-36.⁵

We acknowledge two important limitations in this study. First, as a hospital-based study, its findings may be difficult to generalise to the community. Second, the FDF Composite Score has not been previously validated in a separate study, which is a critical limitation. The absence of validation means the score's reliability and accuracy in consistently categorising the true clinical severity of priapism have not been independently verified.

In conclusion, Nigerian adults with SCA-related priapism were found to have a low HRQoL across all domains of the RAND 36-Item Health Survey, a finding that worsened with increasing severity of priapism. We recommend that HRQoL be prioritised at all healthcare settings involved in managing these patients, to assist them in attaining a meaningful life free from physical, social, emotional, and psychosocial challenges. Additionally, we strongly recommend future validation studies for the FDF Composite Score to confirm its utility and applicability for the clinical categorisation of SCA-related priapism in diverse patient populations.

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Conflict of interest: None to be declared.

Authors' Contributions

I. Abdulqadir handled the manuscript writing, data analysis, and correspondence, and gave final approval of the manuscript. T. Aliyu was responsible for the study design, data collection, data analysis, and manuscript review. A. G. Kuliya, S. G. Ahmed, D. H. Gwarzo, A. A. Yusuf, and I. I. Musa supervised the study and approved the manuscript. M. M. Isyaku, J. U. Suleiman, I. Bishir, and N. Y. Ado carried out data analysis and manuscript review. All authors read and approved the final version of the manuscript.

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