

Prevalence and Clinical Determinants of Priapism Among Male Children With Sickle Cell Disease Seen at University of Benin Teaching Hospital Benin City

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Abstract

Background

Priapism is a serious urological complication of sickle cell disease (SCD) that can lead to permanent erectile dysfunction if not promptly managed. Despite its clinical significance, data on the prevalence and clinical correlates of priapism among male children with SCD in Nigeria are limited.

Objective

To determine the prevalence, clinical pattern and risk factors of priapism among male children with SCD attending the university of Benin Teaching Hospital.

Methods

A cross-sectional study was conducted among male SCD children at Haematology clinic of UBTH. Sociodemographic, clinical and haematologic data were collected using structured questionnaires and medical records. Prevalence of priapism was calculated and participants were categorized based on the presence or absence of priapism. Associations with age, Packed cell volume (PCV), socioeconomic status and hydroxyurea use were analysed using independent t-tests, Chi-square tests and logistic regression. Statistical significance was set at $p < 0.05$.

Results

Out of 120 male children with SCD, 16 experienced priapism yielding a prevalence of 13.3% (95% CI: 8.3–20.6%). Patients with priapism were significantly older (mean 13.4 ± 3.2 years) and had lower PCV ($23.4 \pm 2.8\%$) compared to those without priapism (10.1 ± 4.4 years, $25.9 \pm 2.6\%$; $p < 0.01$). Adolescents (≥ 12 years) and patients with PCV $< 24\%$ had higher odds of priapism (AOR = 3.81, 95% CI: 1.02–14.1; AOR = 4.72, 95% CI: 1.51–14.8, respectively). Most episodes occurred in the early morning (75%) and lasted ≥ 4 hours (56%). Hydroxyurea use was not independently protective likely due to late initiation.

Conclusion

Priapism affects approximately one in eight male children with SCD at UBTH with adolescence and low PCV being major risk factors. Early identification at high-risk patients, prompt medical intervention and preventive strategies including early hydroxyurea therapy are recommended to reduce complications.

Introduction

Sickle cell disease (SCD) is a haemoglobinopathy with associated chronic haemolysis with more than 300,000 children born annually on Nigeria.¹ Episodic vaso-occlusion at the post-capillary venules by sickled red cells is the hallmark of the pathophysiology of this condition. These pathophysiological processes are responsible for the wide spectrum of acute and chronic complications observed in individuals with SCD, including pain crises, stroke, acute chest syndrome and various urogenital complications.^{2,3}

Priapism is a persistent (lasting more than 4 hours) painful erection of the penis occurring with or without sexual stimulation, the condition was first defined by Tripe in 1845 and is named after Priapus- the Greek god of fertility^{4,5}. The majority of cases occur in sickle cell disease (SCD); however, other disease conditions with hyperviscosity may also cause priapism. Two types of priapism are clinically relevant i.e the ischemic/veno- occlusive (low- flow) type and the non-ischemic/arterial(high-flow) type⁶. The high-flow type is due to the formation of arterio-sinusoidal shunts which feed the cavernosa and circumvent the restrictive arteriolar vaso-constriction while the low-flow type is due to sinusoidal thrombosis with veno-occlusion not involving the cavernosal blood flow. Priapism is predominantly of low-flow type in SCD and results from venous outflow obstruction caused by sickled RBCs within the corpora cavernosa^{7,8}.

Nitric oxide (NO) is a critical molecule in the initiation and regulation of normal erection. NO acts through the activation of guanylate cyclase to stimulate cyclic guanosine monophosphate(cGMP) production, which activates protein kinase G. The final result of these events include; blood vessel dilation and penile smooth muscle relaxation. NO induced cGMP also regulates the expression of phosphodiesterase type 5(PDE-5), the key enzyme associated with detumescence by degrading cGMP. The expression of PGE-5 is directly correlated with the tonic level of cGMP⁹⁻¹¹. Thus, a low PDE-5 is directly correlated with the tonic level of cGMP. Thus, a low PDE-5 level is associated with abnormal detumescence. Chronic haemolysis in SCD results in endothelial NO depletion, associated with decreased cGMP level and corresponding low level of PDE-5 which cumulatively leads to an inability to terminate an erection, the chronically low PDE-5 expression in the penile tissues fails to terminate the erection. Thus, the primary working hypothesis for priapism in SCD is related to NO depletion associated with PDE-5 dysregulation⁹. Several genetic markers have been implicated in the pathophysiology of priapism in SCD. Klotho, a membrane protein that regulates NO synthase, has been associated with priapism in SCD. Another marker singled out as the most potent trigger of priapism is integrin, a subunit of the α - β 3 endothelial adhesion molecule that is believed to bind sickle erythrocytes to the endothelium via intermediary protein thrombospondin, von Willebrand factor and ICAM -7¹⁰.

Infection, full bladder when going to sleep and dehydration, trauma has been associated with onset of priapism but there is no yet well-established mechanistic relationship. Some drugs such as anti-depressants, antipsychotics, alcohol, methylphenidate, cocaine have been reported to cause priapism¹⁰⁻¹⁴. It has been reported more common among patients with vasculopathy (recurrent vaso-occlusive crises, acute chest syndrome, leg ulcer, stroke, pulmonary hypertension, nephropathy). Complications

associated with priapism range from clinical and medical to psychological. Physical complications include penile gangrene, severe erectile dysfunction and penile deformity, infertility. Psychological complications include; anxiety, depression, social isolation, stigmatization¹⁵⁻¹⁷.

Although priapism is more frequently discussed in adult SCD populations, evidence suggests that the condition often begins in childhood or adolescence, with early episodes predisposing affected individuals to recurrent attacks and long-term complications such as erectile dysfunction. The reported prevalence of priapism among males with SCD varies widely across studies, ranging from 2% to over 35% depending on age group, study design and geographical locations¹⁷. In paediatric population, despite these devastating complications, priapism is underreported and under-recognized serious complication of SCD among males, with no evidence based primary prevention. The available figures may underestimate the true burden due to under-recognition, delayed presentation, cultural stigma and lack of routine screening in children. This study aimed to assess the prevalence of priapism in SCD male children and factors associated with it.

Aims and Objectives

General aim: To determine the prevalence and identify the clinical determinants of priapism in male children with SCD.

Specific objectives

1. To estimate the prevalence of priapism in male children with SCD attending sickle cell clinic
2. To describe clinical characteristics of priapism (age at first episode, frequency and duration)
3. To identify clinical and laboratory factors associated with a history of priapism (age at diagnosis, steady state Hb, hydroxyurea Use)

Methodology

The study was a hospital-based descriptive cross-sectional study. It was conducted in the Paediatric sickle cell out- patient clinic of the University of Benin Teaching hospital.

Study Population

All male sickle cell disease children aged 1-18 years that attended the clinic between July and September 2025 were consecutively recruited.

Sample Size

All eligible cases that meet the inclusion criteria during the study period were consecutively recruited (total population sampling).

Ethical approval/ consideration

Ethical approval was sought and obtained from the Ethics Committee of University of Benin Teaching hospital Benin city (NHREC -UBTH- HREC/24/12/2022B).

Informed consent and Assent

The information and consent were given to parents/guardians to read and sign. However, written and/or verbal informed assent were obtained from children seven years and above.

Methods

Two research assistants (1 registrar and 1 house officer) were trained to assist in this study with administration of the questionnaires.

The patients and their parents/caregivers were given clear and adequate information in English and/or Bini Languages and allowed to make an informed decision.

Data Collection Procedure

Research assistants trained for the process administered questionnaires privately to ensure confidentiality. Questionnaire contained patient's demographics, SCD history and priapism history and health care utilization.

Data Analysis

The data were entered into the Excel spreadsheet. IBM Statistical Package for Social Sciences (SPSS) version 26.0 was used to analyse the data. Descriptive statistics were used to summarize the sociodemographic, clinical and haematologic characteristics of the participants. Continuous variables such as age, PCV were presented as means and standard deviations. Categorical variables such as socioeconomic status, timing of priapism episodes, duration of episodes, treatment modalities and hydroxyurea use were summarized as percentages and frequencies. The prevalence of priapism among male SCD patients was calculated as the proportion of participants who had experienced priapism relative to the total number of male patients studied. The 95% CI for the prevalence was estimated using the Wilson method. To compare characteristic between patients with priapism and those without priapism, inferential statistical test was applied. The independent samples t- test was used to compare the means of continuous variables such as age and PCV between the two groups. For categorical variables, the Chi- square test was used to assess associations. Bivariate analysis was performed to determine potential risk factors for priapism. Variables such as age category and PCV levels were analysed using odds ratio with 95% CI to estimate the strength of association. Variables that showed statistical significance in the bivariate analysis were subsequently included in a multivariate logistic regression model to identify independent predictors of priapism. A p-value of <0.05 was considered statistically significant.

Results

Prevalence of Priapism

A total of 120 sickle cell disease males were recruited and 16 of them has had priapism. The prevalence of priapism among male SCD children at UBTH was **13.3% (95% CI 8.3–20.6%)**, indicating that approximately 1 in 8 male SCD children experience priapism.

Sociodemographic and Hematologic Characteristics

Patients with priapism were significantly older than those without (mean difference ≈ 3.3 years, $p=0.004$). Patients with priapism had significantly lower PCV levels (mean difference $\approx -2.5\%$, $p<0.001$). In contrast, Socioeconomic status did not show a statistically significant association with priapism, although a higher proportion of those with priapism belonged to the middle socioeconomic class (73.1%). See table I

Clinical Characteristics of Priapism Episodes (n = 16)

The majority of priapism episodes (75%) occurred in the early morning while 25% occurred at night. Nearly 56% of the priapism lasted ≥ 4 hours. Notably 12.5% did not present to the hospital. See table II

Treatment Modalities for Priapism

The most commonly used treatment was medical therapy (37.5%) followed by exchange blood transfusion (25%). 18.7% of patients received no treatment. However, only 1 patient received cavernosal aspiration. See table III

Association between adolescent age and the occurrence of priapism

Among patients aged 12 years and above, 13 cases of priapism were recorded compared with 42 patients without priapism. Adolescents (≥ 12 years) had 4.5 times higher odds of developing priapism compared with younger patients. See table IV

Relationship between severity of Anaemia and the occurrence of priapism

Among patients with PCV $<24\%$, 9 developed priapism while 18 did not. Patients with PCV $<24\%$ had 5.4-fold increased odds of priapism. See table V

Pattern of Hydroxyurea Therapy

Of the 16 patients who experienced priapism, 11 (68.8%) were receiving Hydroxyurea. However, only 54.5% started the Hydroxyurea before priapism occurred. See table VI

Multivariate Logistic Regression (Model)

After adjustment, age ≥ 12 years remained a significant predictor of priapism. Similarly, PCV $<24\%$ remained a strong independent predictor, with affected patients having 4.72 times higher odds of

developing priapism. In contrast, hydroxyurea use was not significantly associated with priapism. See table VII

Prevalence = $\frac{16}{120} \times 100 = 13.3\%$
95% Confidence Interval (Wilson method)
 $95\% \text{ CI} = 8.3\% - 20.6\%$

Table I: Sociodemographic and Hematologic Characteristics

Variable	Priapism (n=16)	No Priapism (n=104)	p-value
Age (years) mean ± SD	13.4 ± 3.2	10.1 ± 4.4	0.004*
PCV (%) mean ± SD	23.4 ± 2.8	25.9 ± 2.6	<0.001*
Middle SES	56.3%	73.1%	0.18
Upper SES	25.0%	17.3%	0.42
Lower SES	18.7%	9.6%	0.21

*Independent samples t-test

Table II: Clinical Characteristics of Priapism Episodes (n = 16)

Variable	Frequency	Percentage
Timing		
Early Morning	12	75%
Night	4	25%
Duration		
<4 hours	5	31.3%
4–11 hours	7	43.8%
12–24 hours	2	12.5%
Did not present to hospital	2	12.5%

Table III: Treatment Modalities for Priapism

Treatment	Frequency	%
Medical therapy	6	37.5
Exchange Blood transfusion	4	25.0
Cavernosal aspiration	1	6.3
No treatment	3	18.7
Home remedy	2	12.5

Table IV: Association between adolescent age and the occurrence of priapism

Age ≥12	Priapism	No Priapism	OR (95% CI)	p-value
Yes	13	42	4.52 (1.22–16.7)	0.018
No	3	62	Reference	

Table V: Relationship between severity of Anaemia and the occurrence of priapism

PCV <24%	Priapism	No Priapism	OR (95% CI)	p-value
Yes	9	18	5.38 (1.75–16.5)	0.002
No	7	86	Reference	

Table VI: Hydroxyurea Exposure

Variable	Frequency	%
On Hydroxyurea	11	68.8
Started after priapism	6	54.5 of users
Started before priapism	5	45.5 of users

Table VII: Multivariate Logistic Regression (Model)

Variable	Adjusted OR	95% CI	p-value
Age ≥12	3.81	1.02–14.1	0.046
PCV <24%	4.72	1.51–14.8	0.008
Hydroxyurea use	0.81	0.26–2.51	0.71

Discussion

The prevalence of priapism observed in this study was 13.3% (95% CI: 8.3–20.6%), indicating that approximately one in eight male SCD children had experienced priapism. This prevalence is consistent with findings from several African studies that report priapism rates ranging between 10% and 40% among male SCD patients^{17,18}. A Nigerian study by Adeyoju et al. reported that approximately 35% of adult male SCD patients had experienced at least one episode of priapism, highlighting the significant burden of this complication¹⁷. Similarly, a study in Ghana documented a prevalence of about 25% among male SCD patients which is higher than the prevalence documented in the present study, suggesting that priapism remains a common complication across West African populations^{19,20}. Differences in prevalence between studies may be attributed to variations in study population, age distribution, diagnostic criteria, and healthcare-seeking behaviour. The relatively lower prevalence observed in the present study compared with some adult-based studies may be explained by the predominantly paediatric population included in this study. Priapism tends to become more frequent with increasing age, particularly during adolescence and early adulthood, due to hormonal changes and progressive vascular damage associated with SCD²¹. Consequently, studies that include more adult participants often report higher prevalence rates.

The clinical characteristics of priapism episodes observed in this study revealed that most episodes occurred in the early morning (75%), while the remaining occurred during the night. This pattern is consistent with the phenomenon of nocturnal penile tumescence, a physiological process that occurs during rapid eye movement (REM) sleep²². In patients with SCD, the abnormal regulation of penile blood flow during these episodes may lead to prolonged erections. Similar findings have been documented in several studies where priapism episodes were reported to occur predominantly during sleep or early morning hours^{17,20,23}.

Regarding the duration of episodes, more than half of the episodes lasted four hours or longer, indicating a significant risk for ischemic priapism and potential erectile tissue damage. Prolonged priapism is considered a urological emergency because ischemia of the corpora cavernosa can lead to fibrosis and subsequent erectile dysfunction if not treated promptly²⁴. The presence of prolonged episodes in this study highlights the importance of early recognition and timely medical intervention. An important observation in the present study was the delay in seeking hospital care among some patients, as approximately 12.5% of affected individuals did not present to the hospital during their episodes. This may reflect lack of awareness among caregivers, stigma associated with genital symptoms, or limited

access to healthcare facilities. Similar delays have been reported in other African studies where patients often resort to home remedies before seeking medical attention ²⁵.

The treatment patterns observed in this study also revealed notable gaps in management. Although some patients received medical therapy or blood transfusion, only one patient underwent cavernosal aspiration, which is considered a standard intervention for ischemic priapism. The low rate of cavernosal aspiration suggests possible limitations in emergency management practices, including lack of specialized expertise or delayed presentation to healthcare facilities. Current clinical guidelines recommend early aspiration of corporal blood combined with intracavernosal sympathomimetic agents for the management of ischemic priapism ²⁴. Failure to implement these interventions promptly may result in poor outcomes, including erectile dysfunction. These findings therefore underscore the need for improved clinical protocols and healthcare provider training in the management of priapism among SCD patients.

One of the major findings of this study was the significant association between age and priapism occurrence. Patients with priapism were significantly older than those without priapism (mean age 13.4 ± 3.2 years vs 10.1 ± 4.4 years, $p = 0.004$). Furthermore, adolescents aged 12 years and above had approximately 4.5 times higher odds of experiencing priapism compared with younger children. This finding is consistent with previous studies which demonstrate that the risk of priapism increases during adolescence, coinciding with the onset of puberty and increased nocturnal penile tumescence ²⁶. Similar findings have been reported in Nigerian studies. For example, Akinyanju and colleagues observed that priapism was more common among adolescents and young adults with SCD, emphasizing the role of hormonal and physiologic factors in its pathogenesis ²⁶. Additionally, research conducted in Jamaica by Emond et al. demonstrated that the incidence of priapism increased markedly after puberty, suggesting that hormonal influences may contribute to the dysregulation of penile vascular tone in SCD patients ²¹. Increased testosterone levels during puberty may enhance erectile responses, while the abnormal nitric oxide signalling seen in SCD predisposes to prolonged erections ²⁷.

Another important finding in the present study was the significant association between low packed cell volume (PCV) and priapism. Patients who experienced priapism had significantly lower mean PCV values compared with those without priapism ($23.4 \pm 2.8\%$ vs $25.9 \pm 2.6\%$, $p < 0.001$). In addition, patients with $PCV < 24\%$ had more than fivefold increased odds of developing priapism. After adjusting for confounding factors in multivariate analysis, low PCV remained an independent predictor of priapism. This finding supports the hypothesis that severe anaemia contributes to the pathogenesis of priapism in SCD. Chronic haemolysis leads to reduced nitric oxide bioavailability due to increased plasma haemoglobin and arginase activity, resulting in endothelial dysfunction and impaired regulation of penile blood flow ^{28,29}. The resulting imbalance between vasodilation and vasoconstriction may predispose to prolonged erection and venous outflow obstruction. Several studies have reported similar associations between anaemia severity and priapism. In a multicentre study involving SCD patients in the United States, Nolan et al. observed that lower haemoglobin levels were associated with an increased risk of

priapism episodes, suggesting that haemolysis-related vasculopathy plays a major role in its development²⁷. Likewise, Burnett and Broderick highlighted the importance of haemolysis-induced nitric oxide depletion as a key mechanism underlying priapism in SCD^{20,28}.

Socioeconomic status (SES), however, was not significantly associated with priapism in the present study. Although a slightly higher proportion of patients without priapism belonged to the middle socioeconomic class, the difference was not statistically significant. This suggests that biological factors may play a more prominent role than socioeconomic determinants in the development of priapism. Similar findings have been reported in other studies, where SES was not identified as a major predictor of priapism among SCD patients^{29,30}. Nevertheless, socioeconomic factors may still influence health-seeking behaviour and access to timely treatment, which could affect outcomes.

Hydroxyurea therapy is widely used in the management of SCD due to its ability to increase foetal haemoglobin levels and reduce vaso-occlusive complications^{31–33}. In the present study, a substantial proportion of patients with priapism (68.8%) were receiving hydroxyurea therapy. However, more than half of these patients started hydroxyurea after the onset of priapism, indicating that the drug was often initiated as a reactive rather than preventive intervention.

The multivariate analysis showed that hydroxyurea use was not independently associated with a reduced risk of priapism in this study. This finding may be explained by the late initiation of therapy, which may limit its preventive potential. Several studies have suggested that early initiation of hydroxyurea may reduce the frequency of priapism episodes by improving haemoglobin levels and reducing haemolysis³⁴. A study by Anele et al. reported that hydroxyurea therapy significantly reduced the recurrence of priapism in SCD patients²³. Therefore, the timing of initiation may be critical for achieving optimal preventive benefits.

The occurrence of complications such as acute chest syndrome, vaso-occlusive crisis, and stroke among patients with priapism further highlights the systemic nature of vascular dysfunction in SCD. Priapism is often considered part of a broader spectrum of haemolysis-associated complications, which include pulmonary hypertension, leg ulcers, and cerebrovascular disease²⁷. The presence of neurologic complications such as stroke in some patients suggests that severe vasculopathy may underlie both priapism and other systemic manifestations of SCD^{34,35}.

Overall, the findings of this study emphasize that age and severity of anaemia are key predictors of priapism among male SCD patients. Adolescents and patients with low PCV appear to be at particularly high risk and may benefit from closer monitoring and early preventive interventions. In addition, the observed delays in hospital presentation and suboptimal treatment practices highlight the need for improved patient education, caregiver awareness, and healthcare provider training.

Conclusion

Priapism remains an important and potentially debilitating complication among male patients with sickle cell disease (SCD). In this study conducted at the University of Benin Teaching Hospital, the prevalence of priapism among male SCD patients was 13.3%, indicating that approximately one in eight patients had experienced this complication. This finding highlights that priapism continues to be a clinically relevant problem in this population and requires increased attention from both clinicians and caregivers.

The study demonstrated that age and severity of anaemia were the most significant correlates of priapism. Patients who experienced priapism were significantly older than those without priapism, and adolescents aged 12 years and above had markedly higher odds of developing priapism. This finding supports previous evidence that the risk of priapism increases during adolescence, likely due to hormonal influences, increased nocturnal penile tumescence, and progressive vascular changes associated with sickle cell disease.

Another key finding was the strong association between low packed cell volume (PCV) and the occurrence of priapism. Patients with PCV levels below 24% had significantly increased odds of developing priapism, even after adjustment for other variables. This observation underscores the role of severe anaemia and haemolysis in the pathophysiology of priapism, which may contribute to endothelial dysfunction and impaired regulation of penile blood flow.

The study also revealed important clinical patterns of priapism episodes. Most episodes occurred during the early morning hours, consistent with the physiologic phenomenon of nocturnal penile tumescence. Furthermore, a substantial proportion of episodes lasted four hours or longer, placing affected patients at risk of ischemic injury to the penile tissue and potential long-term complications such as erectile dysfunction.

Despite the clinical significance of this condition, the findings also revealed suboptimal health-seeking behaviour and treatment practices. Some patients did not present to the hospital during episodes, while others relied on home remedies. In addition, only a small proportion of patients received guideline-recommended interventions such as cavernosal aspiration, suggesting gaps in emergency management and possible delays in presentation to healthcare facilities.

Although a considerable number of patients were receiving hydroxyurea therapy, the drug was often initiated after the occurrence of priapism, indicating that its use was largely reactive rather than preventive. Consequently, hydroxyurea use was not found to be independently protective against priapism in this study. Early initiation of hydroxyurea therapy may therefore be important in reducing haemolysis and preventing complications associated with sickle cell disease.

In summary, this study demonstrates that priapism is relatively common among male SCD patients and is strongly associated with adolescence and severe anaemia. The presence of prolonged episodes and gaps in treatment highlights the need for improved patient education, early recognition of symptoms, and prompt medical intervention. Strengthening preventive strategies, including optimal hematologic

management and early initiation of disease-modifying therapies, may help reduce the burden of priapism and improve the overall quality of life of affected patients.

Recommendations

1. Male SCD patients particularly adolescents aged 12 years and above should be closely monitored for symptom of priapism during routine clinic visits as early detection may help prevent complications
2. Efforts should be made to optimize haematologic status in males with SCD. Regular monitoring of PCV levels along with appropriate interventions such as nutritional support, folate supplementation and transfusion when indicated may help reduce the risk of severe anaemia and related complications
3. There is need for improved caregiver/patient education regarding priapism as a medical emergency. They should be informed about the warning signs of priapism, the importance of early hospital presentation and the potential complications of delayed treatment including erectile dysfunction.
4. Hydroxyurea should be considered early in eligible SCD patients, particularly those at higher risk for complications such as recurrent vaso-occlusive crises or priapism.
5. Health workers should be trained on the standard guideline for the management of priapism, including timely diagnosis and prompt interventions such as cavernosal aspiration and appropriate pharmacologic therapy. Establishing clear clinical protocols in emergency departments may improve treatment outcomes.

Limitations

1. A single tertiary health care centered study may not be fully generalizable to all male sickle cell disease children.
2. The study relied partly on self- reported history of priapism episodes which may introduce recall bias.
3. Although the study examined hydroxyurea exposure, it did not assess duration of therapy, adherence to treatment or dosing levels which may influence its effectiveness in preventing complications.

Declarations

Consent for publication:

Written informed consent was obtained from all individual participants (or their legal guardians) for publication of this study and its accompanying data.

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Author Contribution

Odunvbun Magdalene: conceptualization, study design, data analysis and interpretation, drafting of the manuscript, reviewing of the manuscript
Ezeuko Lilian: conceptualization, study design, data analysis and interpretation, drafting of the manuscript, reviewing of the manuscript
Odunvbun Williams: study design, literature review, data interpretation, revision of the manuscript
Ezeuko Vitalis: study design, literature review, data interpretation, revision of the manuscript

Data Availability

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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