

# Priapism in Children with Sickle Cell Disease: Description of a Series 28 Cases at CHU Conakry

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**Abstract:** *Introduction:* the objective of this study was to describe the epidemiological, clinical, therapeutic and evolutionary aspects of priapism in children with sickle cell disease. *Methodology:* this was a descriptive retrospective study, lasting 6 years from January 1, 2017 to December 31, 2022, carried out at the pediatric surgery unit of the Ignace Deen national hospital and at the SOS center drepa of morykanteyah. It covered 28 records of sickle cell patients aged 0 to 15 years. *Results:* we recorded 28 cases of priapism out of 1050 sickle cell patients, a frequency of 2.66%. The average age of patients was 9.79 years with the extremes of 5 and 14 years, 54% of cases were homogygous, pain and rigidity of the penis were the most frequent clinical signs, i.e. 100%. The duration of symptom evolution was greater than 36 hours in 53.57%. It was an acute priapism in 54% with a nocturnal occurrence in 57%. 39.29% had suffered 1 to 2 episodes of priapism in their history. The medical treatment involved analgesics, rehydration and oral etilefrine in 85.71%, 32.14% and 21.43% respectively. Intracavernous injection of etilefrine was the most used method with 35.71% success rate, while bilateral incision of the corpora cavernosa (distal Al-Ghorab shunt) was the ultimate remedy with 100% success rate. Detumescence was obtained the same day in 60.71%. Fibrosis of the corpora cavernosa and relapse were the complications found. *Conclusion:* priapism in children with sickle cell disease is a relatively frequent urological emergency, early consultation and adequate care could reduce complications.

**Keywords:** Priapism, Sickle Cell Disease, Child

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## 1. Introduction

Priapism is a painful and irreducible prolonged erection occurring without any sexual stimulation and not resulting in ejaculation [1]. It is a common urological complication of sickle cell disease, and is often cited as one of the most difficult clinical problems encountered by pediatric urologists [2]. This sickle cell priapism is a low-flow priapism of which there are two clinical forms: acute priapism (AP), as a rule isolated, of prolonged duration, and chronic intermittent priapism (ICP) made up of repeated attacks of priapism

(several times a week) of short duration, usually less than 6 hours [3, 4]. Its diagnosis is obvious, requiring no additional examination and its management aims to prevent complications such as: shortening of the penis, erectile dysfunction and psychological sequelae [5, 6].

Sickle cell priapism accounts for up to 65% of all priapism episodes occurring in children [7, 8]. Its prevalence varies from one continent to another or from one country to another: In the United States, Mantadakis et al. [9] reported a prevalence of 27.5% of priapism in children with sickle cell disease. In Great Britain Adeyoju et al. [10] reported a

prevalence of 37%. In Africa, several studies have been carried out, particularly in Congo, Togo and Senegal, which have reported respective prevalences of 34% [11], 23.6 [12] and 9.3% [13].

The aim of this study was to describe the epidemiological, clinical, therapeutic and evolutionary aspects of priapism in children with sickle cell disease.

## 2. Material and Methods

This was a retrospective study of the descriptive type, lasting 6 years from January 1, 2017 to December 31, 2022, carried out at the Pediatric Surgery Unit of the Ignace Deen National Hospital and at the SOS Drépa center in Morykantéah. It covered 28 records of sickle cell patients aged 0 to 15 years. We proceeded to a systematic recruitment of all files of children meeting our inclusion criteria. Our study variables were epidemiological, clinical, therapeutic and evolutionary. The data was collected from the files of our respondents and mentioned on a pre-established survey sheet for this purpose. They were entered and analyzed using Word, Excel, Power point from the 2013 office pack and Epi Info in version 7.2.1.

## 3. Results

Out of a total admission of 1050 cases, we recorded 28 cases of sickle cell priapism at the pediatric surgery unit of the Ignace DEEN National Hospital, i. e. a frequency of 2.66%. The age group of 6 to 10 years was the most frequent with a proportion of 57.14 followed by that of 11 to 15 years with a proportion of 39.28%.

The youngest patient was 5 years old and the oldest 14 years old with an average age of 9.79 years. 54% of our patients were homozygous SS forms against 46% heterozygous forms.

The main reported reasons for consultation are shown in Table 1. Penile pain and stiffness were the main symptoms.

**Table 1.** Breakdown of patients by reason for consultation.

Reasons for consultation	Effective	Percentage
Penis stiffness	28	100
Pain	28	100
Fever	19	67.86
Dysuria	12	42.86
Urinary burning	3	10.71

The consultation time is shown in Table 2. This delay had exceeded 36 hours for the majority of patients, i. e. 53.57%.

**Table 2** Distribution of patients according to consultation time.

The consultation period	Effective	Percentage
Inf. à 4h	3	10.71
13-36h	8	28.57
Sup. à 36h	15	53.57
No determined	2	7.14
Total	28	100

Acute priapism was the most common type of priapism in 54%, while intermittent priapism was found in 46% of cases.

The time of onset was nocturnal in the majority of cases, i. e. 57% versus 43%.

The majority 39.29% of our patients had suffered 1 to 2 episodes of priapism in their history; 21.43% 3 to 4 episodes while 25% were in their first episode represented in the table below.

**Table 3.** Distribution of patients according to the number of previous episodes.

Number of previous episodes	Effective	Percentage
1 <sup>er</sup> épisode	7	25
1-2 once	11	39.29
3-4 once	6	21.43
Sup. à 4 once	4	14.28
Total	28	100

Analgesics/antipyretics and nonsteroidal anti-inflammatory drugs were used with a common proportion of 85.71%. In addition, oral rehydration and etilefrine were also administered with 32.14% and 21.43% respectively. Ice pack was applied in 11 patients and had no effect. Puncture-aspiration was performed in 4 patients with detumescence obtained in 2 patients, i. e. 50%. Intracavernous injection of etilefrine was performed in 26 patients with detumescence in 11 patients, a success rate of 35.71%. Bilateral incision of the corpora cavernosa (distal Al-Ghorab shunt) was the last resort in cases of failure of the previous techniques and was performed in 15 patients with detumescence obtained in 100% of cases.

Detumescence was obtained on the same day in 60.71% of patients then in 2 to 3 days in 28.57%. However, a late detumescence of more than 3 days was observed in 3 patients, i. e. 10.71%.

Complications could only be assessed in 10 patients or 35.74% against 18 or 64.28% lost to follow-up. Among the 10 patients, 5 or 50% had a normal erection, 4 or 40% presented with fibrosis of the corpora cavernosa and a relapse or 10%. They are represented in the table below.

**Table 4.** Distribution of patients according to progression.

Organ function	Effective	Percentage
Normal erection	5	50
Fibrosis of the cavernous	4	40
Relapse	1	10
Lost view	18	64.28
Total	28	100

## 4. Discussion

Priapism is a prolonged erection for more than 6 hours in the absence of any sexual stimulation, despite an orgasm [14]. It is a rare urological emergency in children, leading to sexual impotence due to secondary fibrosis of the corpora cavernosa [15]. Priapisms due to cavernous thrombosis are the most common and their main etiology in children is sickle cell disease [15]. Indeed, 5% of children with sickle cell disease are at risk of developing priapism, which is due to an aggregation of red blood cells following crystallization of hemoglobin S [15]. In this study we noted 2.66%. This result

underlines the relative frequency of this complication in sickle cell patients in our context.

The average age noted in this series confirms the data in the literature, which stipulate that the average age in sickle cell endemic areas is less than 15 years [2, 9, 12].

The diagnosis is obvious clinically in view of a stretched penis in irreducible erection, pressed against the abdominal wall. Physical examination shows induration of the corpora cavernosa extending from the perineum to their ischio-pubic insertion. The corpus spongiosum and the glans are not affected [16]. Indeed, penile rigidity, a classic sign of priapism, and pain were found in all our patients. While voiding disorders were dominated by dysuria. Latoundji S. [16] reported in his series that in addition to pain, voiding disorders such as urinary burning were present in all his patients.

Sleep, fever and vaso-occlusive crises are the main predisposing factors found in the literature. During sleep, erections occur but also a slowing of the circulation with a drop in the partial pressure of oxygen [11, 16, 17]. In our series fever was observed in many of our patients.

The consultation time is of great importance, because it determines the functional prognosis of the patient. It implies the pre-therapeutic delay. For the majority of authors [18-20], it should not exceed 36 hours to minimize the risk of erectile impotence. This risk would reach 60% after 48 hours and exceed 80% after four days [21]. However in our series there was a delay in the consultation for the majority of our patients. Same observation in the studies of Gbadoe AD et al in Togo [12] which noted an average consultation time of four days and in Senegal [13] which reported that the consultation took place on average between 10 and 72 hours. These results could be explained by the parents' lack of information on the possibility of the occurrence of priapism as a sickle cell complication and the socio-cultural prejudices which, according to certain studies, priapism is considered to be a natural phenomenon, which can occur in any individual throughout life [11, 13].

The predominance of homozygous forms noted in our study was also reported by Latoundji S et al. [15] who found that the homozygous SS forms occupied 75%, while the double heterozygous SC and heterozygous AS were observed in 16.67% and 8.33% respectively. It still indicates the higher susceptibility of homozygous forms to develop priapism compared to heterozygotes.

The large number of acute priapism compared to chronic intermittent could be justified by the fact that episodes of acute priapism are accompanied by pain, which motivates the consultation.

Even if intermittent priapism seems more frequent, it lasts less than acute priapism and it can resolve spontaneously or with small means (tepid water bath, walking, etc.). Thus, intermittent priapism in sickle cell patients results in intermittent swelling of the penis, or sometimes short and painless perineoscrotal swelling: this is why in the latter, in the event of a sickle cell crisis, it is recommended to systematically inspect the penis [22, 19].

Episodes of nocturnal priapism would be linked to the

known physiological phenomenon of nocturnal erections, a phenomenon that would be related to an increase in plasma testosterone [13].

The management of sickle cell priapism depends on the time to management and the severity of the anoxia.

It includes multiple and varied means and methods. There is no consensus on their implementation. However the simple means must be tried first, the medical treatment is enough in the first 4 hours and the surgical techniques must be used as a last resort ranging from the simplest to the most complex.

The low success rate linked to the intracavernous injection of etilefrine in our series could be explained by the late use of this product due to the particularly long consultation time (more than 36 hours) for the majority of our patients, whereas this injection is only effective when used within 4 hours of the onset of priapism.

After this time, the injection must be associated with the puncture to obtain a good result. Bilateral incision of the corpora cavernosa (distal Al-Ghorab shunt) was the last resort in cases of failure of the previous techniques with detumescence obtained in all patients. This technique has been described by several authors with varying results [23-25]. The detumescence obtained on the same day in the majority of our patients converges with that of Kassogu   et al. [1] who obtained detumescence on the same day in 61% of the patients punctured. However, a late detumescence of more than 3 days was observed in 3 patients, i. e. 10.71%. The major issue in the management of priapism is the preservation of erectile function. This risk seems higher when the treatment occurs after 24 hours of progression [1, 13].

The high number of patients lost to follow-up and the insufficiency of certain files have meant that questions have remained unanswered (eg: the exhaustive evaluation of erectile function, the other complications of sickle cell disease associated, etc.).

## 5. Conclusion

Sickle cell priapism is a relatively frequent urological complication in our context. However, it is important to inform and verify the knowledge of this complication in the sickle cell population while insisting on the importance of an early consultation and the existence of a simple and effective treatment when the diagnosis is early.

## Conflict of Interests

The authors declare that this work presents no conflict of interest.

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