



Outcome of Management of Acute Prolonged Priapism in Patients with Homozygous Sickle Cell Disease

Résultat de la gestion des cas de priapisme aigu prolongé chez les patients homozygotes avec la Drépanocytose

F. O. Adetayo

ABSTRACT

BACKGROUND: Priapism is a persistent, often painful, purposeless penile erection, which may or may not be associated with sexual desire, excitement stimulation, or intercourse.

OBJECTIVE: To present the outcome of management of acute prolonged priapism in patients with homozygous sickle cell disease.

METHODS: Fifty-four patients seen over a 20-year period were studied. Of these, 35 were treated surgically with Ebbehøj's cavernosa-glandular shunt while 19 were treated conservatively. The information documented for each patient included age, haemoglobin genotype, duration of priapism before treatment, time of onset, previous history of priapism and possible aetiological factors.

RESULTS: The age range was 2.5–38 years with a mean of 20.56 + 9.33 years. The potency rate in those treated conservatively was 47.37% while it was 70.37% in those treated surgically. The potency rate decreased with increasing duration of priapism before treatment. Those treated within three days had a significantly better outcome than those treated after three days. ($\chi^2 = 4.2986$, $P = 0.038$). The potency rate also decreased with increasing age at onset but there was no statistically significant difference between the potency rate in the age groups.

CONCLUSION: Surgical treatment of acute prolonged priapism may be associated with a higher potency rate compared to conservative treatment. There is an inverse relationship between the potency rate and duration of priapism before treatment and age at onset of Priapism. Acute prolonged priapism is a common cause of impotence in patients with homozygous sickle cell disease. *WAJM 2009; 28(4): 234–239.*

Keywords: Anaemia, Priapism, HbSS Disease, Sickle cell anaemia, impotence.

RÉSUMÉ

CONTEXTE: Le priapisme est une persistante, souvent douloureuse, l'érection du pénis sans but, qui mai mai ou ne pas être associé avec le désir sexuel, la stimulation de l'excitation, ou les rapports sexuels.

OBJECTIF: présenter les résultats de la gestion de priapisme aiguë prolongée chez les patients atteints de drépanocytose homozygote.

MÉTHODES: Cinquante-quatre patients vus au cours d'une 20 - période d'un an ont été étudiés. De ce nombre, 35 ont été traités chirurgicalement avec caverneux Ebbehøj's shunt glandulaires tandis que 19 ont été traités de façon conservatrice. Les renseignements documentés pour chaque patient comprenaient l'âge, le génotype de l'hémoglobine, la durée du priapisme avant le traitement, le temps d'apparition, les antécédents de priapisme et d'éventuels facteurs étiologiques.

RÉSULTATS: La tranche d'âge était 2.5-38 ans avec une moyenne de 20,56 + 9,33 années. Le taux d'activité dans ces traités de façon conservatrice a été 47,37% alors qu'il était 70,37% chez ceux traités chirurgicalement. Le taux d'activité diminue avec l'allongement de la durée du priapisme avant le traitement. Ceux traités dans les trois jours a eu un résultat nettement meilleur que ceux traités au bout de trois jours. ($\chi^2 = 4,2986$, $p = 0,038$). Le taux d'activité a également diminué avec l'augmentation de l'âge au début, mais il n'y avait pas de différence statistiquement significative entre le taux d'activité dans les groupes d'âge.

CONCLUSION: Le traitement chirurgical de priapisme aiguë prolongée mai être associés à un taux supérieur à la puissance par rapport à un traitement conservateur. Il existe une relation inverse entre le taux d'activité et la durée du priapisme avant le traitement et l'âge d'apparition de priapisme. Priapisme prolongé aiguë est une cause fréquente d'impuissance chez les patients atteints de la maladie drépanocytaire homozygote. *WAJM 2009; 28(4): 234–239.*

Mots-clés: Anémie, priapisme, HbSS Disease, Sickle cell anaemia, l'impuissance

Urology Unit, Department of Surgery, College of Medicine, University of Lagos, Idi-Araba, Lagos, Nigeria

*Correspondence: Dr F. O. Adetayo.

Abbreviations: APP, Acute prolonged priapism; HbSS, Sickle haemoglobin; HbSS, Homozygous sickle cell disease.

INTRODUCTION

Priapism is defined as persistent often painful purposeless penile erection which may or may not be associated with sexual desire, excitement, stimulation or intercourse.

It was first described by Tripe in 1845 but its association with homozygous sickle cell disease was first recognized by Diggs and Chings in 1934.¹

Sickle Haemoglobin (HbS) is an example of a single point mutation in which the codon determining the amino acid at position six on the β chain ($\beta 6$) has changed from GAG coding for glutamic acid to CTG coding for valine.² Sickle haemoglobin polymerizes when deoxygenated resulting in abnormal sickle shaped erythrocytes that are rigid and can not transverse small capillaries. When this occurs in the corpora cavernosa, there is localized stasis and obstruction of the venous drainage with resultant low-flow priapism. Sickle cell disease refers to various related disorders characterized by the predominance of sickle haemoglobin (HbS) production. The most common of these disorders is homozygous sickle cell disease (HbSS) or sickle cell anaemia. Sickle cell trait describes heterozygous carriers of the HbS gene.

The prevalence of priapism in patients with homozygous sickle cell disease has been reported to be between 33% and 42%.^{3,4}

Five clinical categories of priapism have been described in patients with sickle cell disease viz, recurrent acute priapism, acute prolonged priapism, chronic priapism, tricorniporeal priapism and high flow priapism.^{2,5}

Acute prolonged priapism (APP) describes a painful erection that does not subside after several hours.⁵ If untreated, the attack will last for several weeks resulting in total impotence.⁵

In 1976, Aina from our hospital reported his experience with cavernosaphenous shunt in the treatment of priapism. He obtained a potency rate of 57%. The three patients who were impotent had HBSS disease and their poor result was attributed to the underlying disease.⁶

This prompted us to conduct a comparative study of the surgical

treatment of priapism in patients with HbSS disease (sicklers) and HbAS and HbAA (non-sicklers) using a simpler operative procedure – the Ebbehoj's. Technique of cavernosa-glandular shunt –.^{7, 8}

Direct aspiration of the corpora cavernosa as a treatment modality has been used in our hospital in a limited number of patients and the result was only slightly better of than conservative management.⁹

This article presents the outcome of management of acute prolonged priapism in 54 patients with homozygous sickle cell disease collected over a twenty years period.

SUBJECTS, MATERIALS, AND METHODS

Sixty-one patients with homozygous sickle cell disease presenting with priapism were seen by the Urology unit of Lagos University Teaching Hospital over a twenty year period 1988–2007. Fifty-four of them had acute prolonged priapism or major attacks of priapism (study population) while seven patients had troublesome stuttering attacks of priapism or recurrent acute priapism.

For each patient in the study population, a full history was taken and complete physical examination was carried out including digital rectal examination and blood pressure check. Urine was sent for microscopy, culture and sensitivity. The information documented for each patient included age, haemoglobin genotype, previous history of priapism, time of onset of priapism, duration of priapism before treatment, possible aetiological factors, associated urinary tract signs and symptoms, other associated signs and symptoms, time of complete detumescence and post-therapy erectile function.

All the patients were initially commenced on medical treatment which consisted of intravenous fluids, generous analgesics, sedatives and when indicated blood transfusion. Those who did not respond to these measures within 12 hours were offered surgery. Those who refused surgery were continued on conservative management.

Our favoured surgical procedure

was Ebbehoj technique of cavernosa – glandular shunt. For the surgically treated patients, a small skin incision, about 1cm in length was made on the dorso-lateral aspect of the glans penis, after the patient had been catheterized. Using a size 15 blade, the incision was deepened and carried down to the tunica albuginea of the tip of the corpus cavernosum on the same side. The tip of the corpus cavernosum was punctured and the tip of the scalpel inserted further into the corpus cavernosum. The scalpel was then rotated through 360° to enlarge the shunt between the corpus cavernosum and the glans penis.

Immediately, viscid altered blood was observed to spurt out through the incision. The corpora cavernosa were then massaged from the perineum to the shaft of the penis to completely evacuate the viscid altered blood. The corpora cavernosa were then irrigated with normal saline until the effluent was bright red. If this did not result in immediate complete detumescence, the procedure was repeated on the other side. The skin incision was closed with 3/0 silk or 3/0 nylon.

Post-operatively, the patient was given analgesic, sedatives, and the intravenous fluid was continued for another 24 hours. He was given antibiotics only when indicated. Possible complications were looked for and treated. The catheter was removed when the patient became pain-free.

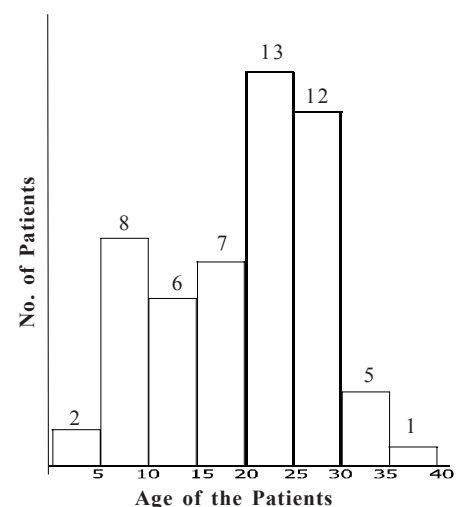


Figure 1: Distribution of Patients with Homozygous Sickle Cell Disease with Acute Prolonged Priapism by Age

Nineteen of the fifty-four patients were treated conservatively while the remaining thirty-five were treated surgically.

One patient had aspiration and irrigation of corpora cavernosa with one in 10⁶ adrenaline in normal saline with transient effect. He subsequently developed a blood pressure of 220/130 mmHg which came back to normal after several hours. He eventually had surgery but failed to return for follow up. Seven other surgically treated patients did not return for follow up.

The assessment of the erectile function was carried out at three months and six months post-therapy. Grading of result was based on the system proposed by Moloney, Elliot and Johnson¹⁰ and is as follows.

- A. Good – when the penis is palpably and functionally normal i.e. good erection with no residual induration of the corpora cavernosa.
- B. Fair – when the patient is able to have useful erection but has some residual fibrotic induration or flaccidity.
- C. Poor – complete inability to achieve useful erection.

Those who were graded A or B were regarded as potent while those who were graded as C were regarded as impotent. The erectile function of those treated conservatively was compared with that of those treated surgically using chi square analysis.

Also, the erectile function of those treated within one day, three days and after three days of onset of priapism was determined, tabulated and subjected to chi-square analysis. Finally, the erectile function of those in the age groups 0-15 years, 16-30 years and those above 30 years were determined, tabulated and subjected to chi square analysis.

The level of significance was fixed at P=0.05 in all cases. The stastical analysis was carried out with SPSS statistical package.

RESULTS

The mean age of the patient was 20.56 + SD 9.33 years and the age range was 2.5 to 38 years. Figure 1 shows the age distribution of the patients in this

Table 1: Treatment Modality in HbSS Patients with Acute Prolonged Priapism.

Age Range (Years)	Number		Total(%)
	Surgical Treatment	Conservative Treatment	
0 – 15	8	8	16(29.6)
16 – 30	24	8	32(59.3)
>30	3	3	6(11.1)
Total	35	19	54(100.0)

study. There are two peak age groups i.e five-ten years and 20-25 years. Majority of the patients (59.26%) were between 16 and 30 years (Table 1). The mean age of those treated conservatively was 20.78 + SD 8.32 years (range 3-35 years) and that of the surgically treated patients was 20.0 + SD 10.45 years (range 2.5-35 years).

There was seasonal variation in the frequency of acute prolonged priapism in this study. Twenty-seven of the 45 patients in which this parameter was recorded occurred during the dry season (November – March) while 18 occurred during the rainy season (April – October). The frequency fell drastically in April and May and then rose steeply and peaked in September.

The time of onset of priapism was at night in 43 (79.6%) patients and during the day in seven (13%) patient. The time of onset was not indicated in four (7.4%) patients. Thirty-eight (70.4%) patients had a previous history of priapism.

Twenty-four (44.4%) of these patients had stuttering attacks of priapism, six (11.1%) had multiple episodes while eight (14.8%) had single episode.

The associated clinical findings in this study were Hepatomegaly in 24 (44.44%) patients, pyrexia in 19 (35.2%) patients and jaundice in 32 (59.26%) patients.

One patient, had heamaturia, low platelet count, hepatosplenomegaly, severe pallor, jaundice, bilateral inguinal and axillary lymphadenopathy. Post-operatively he developed ischaemic area on the penile shaft and scrotum which later ulcerated.

One patient had a markedly enlarged phallus and two others developed enlarged phallus postoperatively. Another patient each had dorsal and ventral chordée.

The treatment received by the patients before they were referred to us included: (a) Intravenous fluid; (b) Analgesics such as pethidine, tramadol,

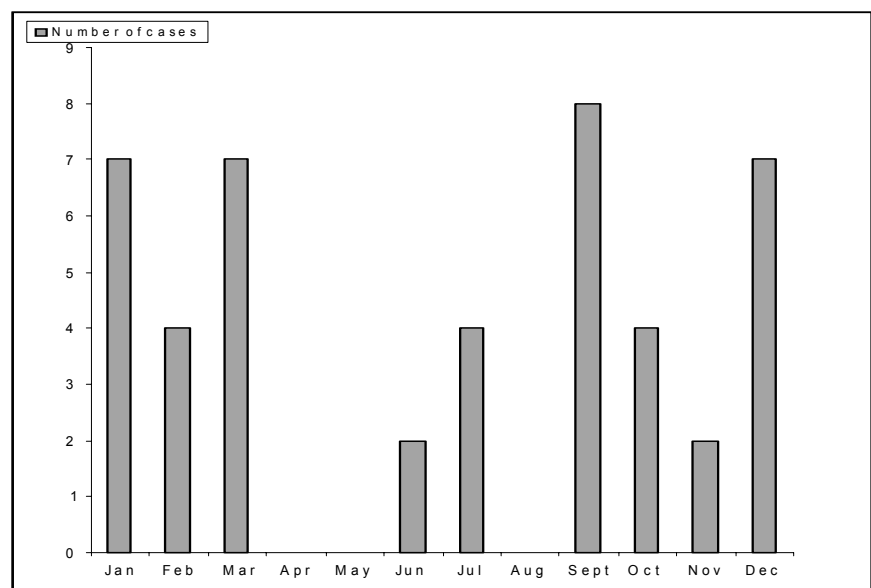


Fig 2. Distribution of Patients with Acute Priapism by Calendar Month

Table 2: Associated Possible Aetiological Factors

Factor	Number
Psychiatric patient	1
Chlorpromazine	2*
Marijuana	2*
Alcohol	3*
Aphrodisiac Tablet	1
Prolonged Sexual Intercourse	2
Diclofenac Sodium and Piroxicam	1
Urethritis	1
Herbal Concoction	1
Thrombosed dorsal vein of the penis	1

*Some of the patients had more than one factor.

pentazocine, diclofenac sodium, brufen and piroxicam. (c) Sedatives such as chlorpromazine, diazepam and bromazepam. (d) Antibiotics such as ceftazidime and gentamycin. (e) Antimalarials such as paludrine. (f) Antihypertensive, such as nifedipine and others like hydroxyurea, ice compress and steroid cream.

Table 2 shows the possible associated aetiological factors.

Two patients developed a recurrence of priapism in the immediate postoperative period. One had repeat

Table 3: Relationship of Potency Rate to Duration of Priapism and Age at Onset

Variable	Potency Rate (N%)	χ^2	P value
SURGICAL TREATMENT			
Duration of Priapism(days)			
< 1	6(100.0)		
< 3	19(86.7)		
> 3	16(50.0)	4.2986	0.038
Age Range (Years)			
0-15	8(85.7)		
16-30	24(70.6)	2.7664	0.251
> 30	3(33.3)		
CONSERVATIVE TREATMENT			
Duration of Priapism(days)			
< 1	5(80.0)		
< 3	10(70.0)		
> 3	9(22.2)	4.337	0.0372
Age Range (Years)			
0-15	8(62.5)		
16-30	84(37.5)	1.284	0.5262
> 30	3(33.3)		

cavernosa-glandular shunt and became impotent. The other refused further surgical treatment and was managed conservatively and retained his potency.

Three other patients had a recurrence of priapism after surgical treatment-one at 13 days, the other, at three months and the last one three years after the first episode. The one, who developed a recurrence at three months, developed another episode three months after the last one. All these recurrences except one were treated surgically, with good result. The one who developed a recurrence 13 days post-surgery was managed conservatively with good result.

Table 3 shows the variation of potency rate with duration of priapism before treatment and age at onset.

Those who were treated within three days of onset of priapism had better results than those who were treated after three days of onset.

The difference in the outcome in the two groups was statistically significant ($\chi^2=4.2986, P=0.038$)

Those who were treated within three days of onset of priapism had comparable outcome irrespective of whether the treatment was surgical or conservative ($\chi^2 = 0.260, P = 0.6098$). However those who were treated after three days had better outcome with surgery compared to conservative treatment ($\chi^2=0.711, p=0.3991$).

The younger patients had better outcome compared to the older ones however the difference was not statistically significant ($\chi^2 = 1.284, p = 0.5262$). Those who were between 0-15 years had comparable result irrespective of whether the treatment was surgical or conservative ($\chi^2 = 0.184; P = 0.6678$). Those who were in the 16 – 30 years age group had better outcome with surgery though the difference was not statistically significant ($\chi^2 = 1.294, P = 0.2552$). Overall, those who were treated surgically had better outcome but the difference was not statistically significant ($\chi^2 = 1.606, p=0.2051$).

DISCUSSION

The incidence of sickle cell disease in Nigeria varies from 0.3% to 2% and that of sickle cell trait is 9 – 20%.¹¹ Each year about 156,000 infants are born with

sickle cell disease of whom 130,000 are in Africa and 33,000 in Nigeria alone.¹² The actuarial probability of young males with homozygous sickle cell disease (HbSS) experiencing priapism by 20 years of age has been estimated to be 89%.¹³ Majority of them occur at night as shown by this study and other studies.^{3,14}

There appears to be a seasonal variation in the occurrence of acute prolonged priapism in our patients (Figure 2). Those occurring during the dry season may be due to dehydration while those occurring during the peak of the rain may be due to malaria and other infections.¹⁵

Marijuana, alcohol, aphrodisiac and prolonged sexual intercourse have been known to potentiate sickle cell disease related priapism.² However diclofenac sodium and piroxicam have not been reported to cause priapism or potentiate sickle cell disease related persistent erection. Chlorpromazine and nifedipine have been associated with priapism and may potentiate a sickle cell disease related episode.¹⁶ They should therefore be avoided.

Priapism is a common complication in patients with sickle cell disease^{3,4} however there is no consistency in the treatment.¹⁷

Conservative measures that have been used include hydration, alkalinisation with sodium bicarbonate, stilboesterol, vasodilators, hydralazine, pentoxifylline, injection of hyaluronidase, controlled hypotension, ice packs, gonadotrophin releasing hormone analogue, hot compresses, sitz baths, anticoagulants, inhalation of 100% oxygen, hyperbaric oxygen, blood transfusion, exchange transfusion, general anaesthesia and spinal, caudal, epidural or pudendal blocks.^{2,4,18,19,20}

Some of these measures are logical on theoretical grounds and some have been occasionally successful while others have been found to be associated with serious complication.²¹ Moreover, many of them have not been assessed in controlled trials.

Many workers have advocated alkalinization to ameliorate the acidosis associated with priapism, this has not proved beneficial.¹⁸ Supplemental oxygen can be administered when

patients are hypoxic with blood oxygen saturation less than 88% but empirical oxygen use in patients without hypoxia may suppress bone marrow stimulation of erythropoiesis.¹⁸

Direct aspiration of the corpora cavernosa through a wide bore needle has been successful in some patients, partially successful in some and failed completely in others.² Recurrence of priapism may necessitate repeat aspiration. Occasionally, aspiration has given rise to serious complications such as infection at the puncture site, sloughing of the prepuce or abscess of the penile shaft, with necrosis of the penis.²

Aspiration can be combined with irrigation with saline, metaraminol, phenylephrine, epinephrine, etilephrine, ephedrine or noradrenaline.^{2,13,19,20} However, clinicians must be aware that some of these alpha - adrenergic agonists have an additional β -one action that can result in systemic adverse events such as severe hypertensive crisis, pulmonary oedema and even death due to rupture of aneurysm.²² Moreover sickle cell pulmonary hypertension, a major risk factor for premature death, affects about 30% of patients with HbSS.²³ Priapism and stroke have been found to be more common in this group of patients.²⁴

The surgical treatment of priapism consists of a shunt operation between the engorged corpora cavernosa and the flaccid corpus spongiosum or the venous system. These include caverno – saphenous shunt,⁶ caverno-spongiosum shunt, caverno-spongiosum shunt with saphenous vein patch,¹¹ corpus cavernosum to dorsal vein of the penis shunt, Winter's cavernosa-glandular shunt, Ebbehøj's cavernosa-glandular shunt and Algorab or modified carvenospongiosum shunt – ^{2,18,19,20,25}.

It has been reported that surgical intervention is associated with a lower average potency rate than medical intervention.¹⁴ It is possible that such surgical patients received delayed treatment causing bias towards failure.^{2,18} In this study those who were treated surgically had higher potency rate (70.37%) compared to those who were treated conservatively (47.37%). All the patients who were treated surgically and

80% of those treated conservatively within 24 hours were potent and the potency rate decreased with increasing duration of priapism before treatment (Table III). This may be due to the fact that priapism lasting for 12 hours causes trabecular interstitial oedema, followed by destruction of sinusoidal endothelium and exposure of basement membrane in 24-hour.²⁶ Formation of sinusoidal thrombi smooth muscle cell necrosis and fibrosis occur in 48 hours.²⁶

The potency rate also decreases with age in both the conservative group and the surgical group. This agrees with the finding of Charkrabarty *et al.*²⁷

Eighteen of our patients were impotent post therapy. This may be due to fibrosis in the corpora cavernosa or persistence of the shunt in the surgically treated patients.^{2, 25}

In conclusion, surgical treatment of acute prolonged priapism in patients with HbSS disease may be associated with higher potency rate compared to conservative treatment. Acute prolonged priapism is a common cause of impotence in patients with homozygous sickle cell disease. Early recognition and prompt treatment offer the best opportunity to avoid this major problem.

Considering the limitations of this study which include possible selection bias and incomplete data especially in the surgically treated group, there is need for well-designed adequately powered multi-centre randomized controlled trials to assess the effectiveness of specific interventions for priapism in patient with sickle cell disease.²⁸

REFERENCE

1. Diggs LW, Ching RE. Pathology of sickle cell anaemia. *South Med J.* 1934; **27**: 839–49.
2. Serjeant, GR (editor) Priapism: In Sickle cell disease 3rd Edition New York: Oxford University Press (2002); 326–338.
3. Kehinde MO, Ibidapo MOO, Okany CC. Priapism in sickle cell anaemia patients at Lagos Tertiary Hospital. *Nig J of Int Med* 2000; **3**: 35–40.
4. Emond AM, Holman R, Hayes RJ, Serjeant G. R. Priapism and impotence in Homozygous Sickle Cell Disease. *Arch Intern Med.* 1980; **140**: 1434–1437.
5. The sickle cell information centre. The Georgia comprehensive sickle cell center

- at Grady Health System Services N. I. H. Guidelines: Management and therapy of sickle cell disease chapt. 19 – Priapism N. I. H. Publication 1995 No 95–2117: 1–3.
6. Aina O. Experience with caverno-sapheous shunt in priapism, West Africa Journal Surgery. 1976; **1**: 59–62.
7. Adetayo FO, Osegbe DN. Priapism; Result of surgical treatment in sicklers and non-sicklers at the Lagos University Teaching Hospital. *The Nigerian Postgraduate Medical Journal* 1994; **1**: 9–12.
8. Ebbehøj JA. New operation for Priapism. *Scand J Plast Reconst Surg.* 1975; **8**: 241–242.
9. Adetayo FO. Management of Priapism in Lagos University Teaching Hospital. *Nig Qt J Hosp Med.* 1997; **7**: 349–353.
10. Moloney PJ, Elliot GB, Johnson HW. Experiences with priapism. *J Urol.* 1975; **114**: 72–76.
11. Odelowo EO. A new caverno-spongiosum shunt with saphenous vein patch graft for established priapism. *Int surg.* 1988; **73**: 130–132.
12. Fleming AF, DeSilva PS. Haematological diseases in the Tropics in Manson-Bahr Wilkoeks (eds) Manson's Tropical Disease Bailliere Tindall 199–208.
13. Mantadakis E, Cavender JD, Rogers ZR, Ewalt DH, Buchanan GR. Prevalence of priapism in children and Adolescents with sickle cell anaemia. *J Paediatr Haematol Oncol.* 1999; **21**: 518–522.
14. Hamre MR, Harmon EP, Kirkpatrick DV, Stern MJ, Humbert JR. Priapism as a complication of sickle cell disease. *J Urol.* 1991; **145**: 1–5.
15. Parry, E. H. O. (editor) Practice of medicine in Africa. 2nd edition Oxford University Press 1984: 1–23 and 753–760.
16. Maclean F, Lee A: Drug – induced sexual dysfunction and Infertility. *The Pharmacological Journal* 1999; **262**: 780–784.
17. Chinegwudoh, F; Anie, K; Treatment of Priapism in boys and men with sickle cell disease. *Cochrane – Database – Ssystem Rev.* 2004 (4) CD004198.
18. Bruno D, Wigfall DR, Zimmerman SA, Rosoff PM, Wiener JS. Complications of sickle cell disease. *J Urol.* 2001; **166**: 803–811.
19. Al Burnett: Pathophybiology of priapism: Dysregulatory erection Physiology Thesis. *J Urol.* 2003; **170**: 26–34.
20. Adetayo FO. Priapism: A review *Nig. Qt J. Hosp. Med* 1998; **8**: 164–169.
21. Siegel JF, Rich MA, Brock WA.

- Association of sickle cell disease, priapism, exchange transfusion and neurological events: ASPEN syndrome. *J Urol.* 1993; **150**: 1480–1482.
22. Ballogiannis DM, Charalabopoulos AK, Giannakopoulos XK, Giannakis DJ, Sofikitis NV, Charalabopoulos KA. Penile erection during transurethral surgery: *Journal of Andrology* 2006; **273**: 1–9.
 23. Gladwin MT, Sachdev V, Jison ML. Schiztukuda Y, Plehn JF, Minter K. *et al.* Pulmonary Hypertension as a risk factor for death in patients with sickle cell disease. *N Engl-J Med* 2004; **350**: 886–895.
 24. Nolan VG, Wyszynski DF, Farrer LA, Steinberg MH. Haemolysis – associated priapism in sickle cell disease. *Blood* 2008; **106**: 9: 3264–3267.
 25. Aghaji AE. Priapism in adult Nigerians B.J.U International 2000; **85**: 493–495.
 26. Reynard J, Brewster S, Biers S (editors) Disorders of erectile function, ejaculation and seminal vesicles: In Oxford Handbook of Urology Oxford University Press First Edition 2006: 488–489.
 27. Chakrabarty A, Upadhyay J, Dhabuwala CB, Sanai K, Perlmutter, AD, Conor JP. Priapism associated with sickle cell haemoglobinopathy in children: long-term effects on potency. *J Urol.* 1996; **155**: 1419–1423.
 28. Maples BL, Hagemann TM. Treatment of Priapism in Pediatric patients with sickle cell disease. *Am J Health System Pharma.* 2004; **61**: 355–363.