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Surgical Management of Stuttering Ischemic Priapism: A Case Report and Concise Clinical Review



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ABSTRACT

Stuttering priapism is an extremely rare and poorly understood entity. We present a rare case of a 47-year-old Afro-Caribbean gentleman who required proximal shunt procedure to treat his ischemic stuttering priapism after he had failed medical management. We provided a concise review of the literature on the surgical management of ischemic priapism. This case highlighted the importance of prompt surgical intervention in prolonged stuttering priapism to avoid serious psychological and functional complications.

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Introduction

Stuttering priapism is an extremely rare and poorly understood entity. It is defined as recurrent episodes of painful and often self-limiting erections lasting <4 h. The etiology of stuttering priapism is similar to that of ischemic priapism [1]. Prolonged stuttering (>4 h) ischemic priapism mandates rapid and compulsory intervention to prevent penile fibrosis and subsequent impotence with a primary aim is to prevent further priapic episodes [2].

Case report

A 47-year-old Afro-Caribbean gentleman presented with 8-h duration of painful priapism. He denied recent use of intra-cavernosal agents, pelvic injury or sexual activity. He attended ED a month ago with similar presentation, which resolved spontaneously. He was found to have sickle cell trait. His past medical history includes allograft kidney transplant for hypertensive end-stage CKD and familial amyloidosis. His medications include Omeprazole, Myofortic, Prograf, Prednisolone, Aspirin, Amlodipine and Doxazosin. Examination revealed engorged corpora cavernosa. Cavernosal blood gas analysis showed PH: 7.24, pO₂ 5.7 kPa, PCO₂ 5.6 kPa, lactate 3.75 mmol/l. This is consistent with ischemic stuttering priapism [2]. Despite 2 emergency cavernosal aspirations and alpha-adrenergic agonist, the patient fails to respond to treatment. Distal shunt procedure was performed to achieve

detumescence. However, priapism recurred within 12 h period and the patient underwent a proximal shunt procedure (corpora-spongiosal shunt).

Discussion

Priapism is a rare symptom with diverse etiological factors. It can be divided into 3 categories according to the pathophysiology; ischemic, nonischemic and stuttering. In this manuscript we discuss the surgical management of ischemic stuttering priapism. The pathophysiology of this type of priapism is not fully understood. A mechanism based on dysregulation of nitric oxide and phosphodiesterase type 5 in the corporal smooth muscle has been proposed [3].

Although most cases in adults are secondary to drug use and intra-cavernosal injections, it also can represent underlying hematologic and metabolic disorders. A list of etiologies is represented in Table 1 [1]. Our patient had multiple risk factors for developing stuttering ischemic priapism, which include race, renal transplant, sickle cell trait and alpha-blocker. There is a well-established evidence associating priapism with sickle cell disease, renal failure and the use of alpha blockers [1,4].

Management of stuttering ischemic priapism should progress in an aggressive and stepwise fashion to achieve prompt resolution. A multidisciplinary approach is often required involving hematologists and renal physicians (particularly in this case in light of the patient's PMH). Treatment of stuttering priapism is primarily medical with an ultimate goal is to prevent further episodes. However in cases, which are refractory to prolonged conservative

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Table 1
Aetiological factors for ischemic stuttering priapism

Idiopathic	
Hematologic dyscrasias	Sickle cell disease, thalassemia, leukemia; multiple myeloma, hemoglobin Olmsted variant, fat emboli during hyperalimentation, hemodialysis, glucose-6-phosphate dehydrogenase deficiency, factor V Leiden mutation
Infections (toxin mediated)	Scorpion sting, spider bite, rabies, malaria
Metabolic disorders	Amyloidosis, Fabry disease, gout
Neurogenic disorders	Syphilis, spinal cord injury, cauda equina syndrome, autonomic neuropathy, lumbar disc herniation, spinal stenosis, cerebrovascular accident, brain tumor, spinal anesthesia
Neoplasms (metastatic or regional infiltration)	Prostate, urethra, testis, bladder, rectal, lung, kidney
Medications	<ol style="list-style-type: none"> Vasoactive erectile agents (Papaverine, Phentolamine, Prostaglandin E1/Alprostadil, combination of intracavernous therapies) Adrenergic receptor antagonists (Prazosin, Terazosin, Doxazosin, Tamsulosin) Antianxiety agents (Hydroxyzine) Anticoagulants (Heparin, Warfarin) Antidepressants and antipsychotics (Trazodone, Bupropion, Fluoxetine, Sertraline, Lithium, Clozapine, Risperidone, Olanzapine, Chlorpromazine, Thioridazine, Phenothiazines) Antihypertensives (Hydralazine, Guanethidine, Propranolol) Hormones (Gonadotropin-releasing hormone, testosterone) Recreational drugs (Alcohol, marijuana, cocaine)

treatment, surgical intervention maybe required to preserve penile structural and functional integrity.

In our case, failed emergency conservative treatment (alpha-adrenergic agonist injection and cavernosal aspirations) has prompted surgical intervention. The goal of surgery is to create a channel or fistula that allows deoxygenated blood to drain from the corpora cavernosum. There are 4 types of shunts [5].

1. Cutaneous distal shunt
2. Open distal shunt
3. Proximal shunt
4. Vein anastomosis/shunts

In this case, distal shunt procedures (Table 2) were ineffective to achieve complete detumescence. A decision therefore was made to perform a proximal shunt procedure (corporospongiosal shunt or otherwise known as Quackels shunt) This is performed in lithotomy position, bulbocavernosus muscle dissected from corpus spongiosum, both corporal bodies incised (12 mm), corporal biopsy was taken and the defects anastomosed together (Fig. 1A and B). Complete detumescence was achieved and patient reported a satisfactory

Table 2
Distal shunt procedures

Winter's procedure	Performed by inserting a large biopsy needle/or cannula through glans into corpora cavernosum several times creating multiple fistulae
Ebbehoj	Performed by percutaneously passing #11 blade scalpel multiple times through glans into corpus cavernosum creating openings in the tunica albugenia resulting in larger fistulae.
T-Shunt (Modified Ebbehoj)	Performed by using no.10 blade scalpel and turning scalpel 90° when pulling out creating 'T-shaped' openings in tunica albugenia

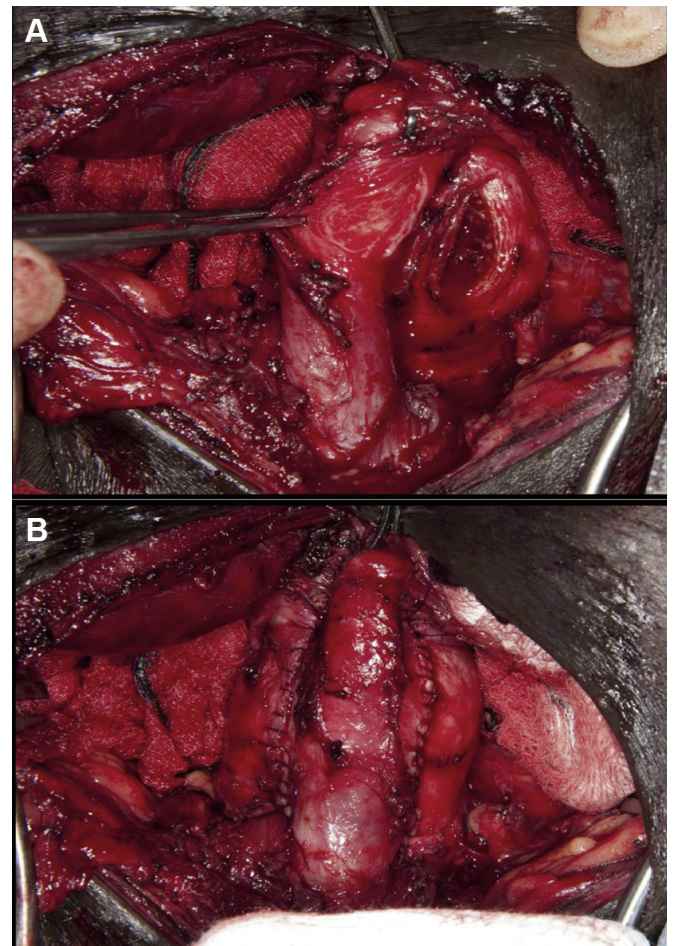


Figure 1. A: Incision in the spongiosum laterally and in the cavernosum medially. B: Anastomosis between corpus cavernosum and spongiosum.

penile function during subsequent clinic follow up. Another proximal shunt operation is Corporo-saphenous vein or superficial/deep dorsal vein shunts (Grayhack or Barry shunt), where one of these veins is ligated and anastomosed with corpora cavernosa.

There is paucity in the literature to favor one shunt procedure over another. However EAU guidelines recommend attempting distal shunt procedures first [1]. As it has lower ED rate <25% compared to 50% with proximal shunt procedure. Priapism patients often receive multiple treatment modalities and it is difficult to ascertain which one has caused the adverse event. Moreover the incidence of ED is proportional to the duration of priapism.

A summary data of case reports and case series generated by American Urology Association showed that the success rates are 66%, 73%, 74%, 76% and 77% for Winter's shunt, Ebbehoj, Al-Ghorab, Grayhack and Quackles respectively [2]. We are aware that further conclusions cannot be reached in the absence of randomized controlled trials.

Conclusion

Stuttering priapism is poorly recognized disorder by many medical professionals. Its management requires multidisciplinary approach with a primary aim is to preserve the penis and to obviate the risk of priapic episodes in the future. This case highlighted the importance of prompt surgical intervention in prolonged stuttering priapism to avoid serious psychological and functional complications.

Conflicts of interest

There is no conflict of interest.

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