ORIGINAL RESEARCH—PRIAPISM

Penile Doppler Ultrasound in Men with Stuttering Priapism and Sickle Cell Disease—A Labile Baseline Diastolic Velocity Is a Characteristic Finding

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ABSTRACT

Introduction. Stuttering priapism (SP) is seen in sickle cell disease (SCD) and characterized by short-lived painful erections. Imbalanced vascular tone is the postulated cause and this may be reflected in changes in baseline penile blood flow as measured using penile Doppler ultrasound (PDU).

Aim. The aim of this study was to investigate the baseline penile blood flow characteristics in men with SCD and SP, by comparing with men without SP.

Methods. PDU findings were retrospectively analyzed in 100 men during flaccid state. Nine men had SP (age range 20–40 years), 4 had Peyronie's disease (PD) (35–48 years), 67 men had erectile dysfunction (16–67 years), and 20 men had normal erectile function (18–42 years).

Main Outcome Measures. The variables measured were peak systolic and end-diastolic velocities, and the Doppler velocity waveform. Values in men with SP were compared with those in the other groups.

Results. Median systolic and diastolic velocity was significantly higher in men with SP (systolic/diastolic velocity was 26/4 cm/second in men with SP vs. 13/0 cm/second, 14/0 cm/second, and 16/0 cm/second in men with PD, ED, and normal erectile function, respectively; $P = 0.0001$). Men with SP had a characteristic low peripheral resistance (PR) waveform with fluctuating velocities; the diastolic velocity was consistently positive (2–7 cm/second) and fluctuated between +2 and +8 cm/second. In comparison, the other 91 men had high PR waveform and consistently negative diastolic velocity (range 0 to –2 cm/second).

Conclusions. Men with SP had a unique baseline Doppler ultrasound waveform, with a low PR waveform and an elevated, variable cavernosal artery velocity. We propose that this may be the sonographic manifestation of a reduced, fluctuating smooth muscle tone and that PDU may have a role for diagnosis and therapeutic monitoring of SP. **Patel U, Sujenthiran A, and Watkin N. Penile Doppler ultrasound in men with stuttering priapism and sickle cell disease—A labile baseline diastolic velocity is a characteristic finding. J Sex Med 2015;12:549–556.**

Key Words. Stuttering Priapism; Sickle Cell Disease; Penile Doppler; Ultrasound

Introduction

Priapism is a penile erection that continues hours beyond or is unrelated to sexual simulation and has traditionally been classified into low-flow (ischemic, veno-occlusive), high-flow (non-ischemic, arterial), or stuttering priapism (SP). SP is characterized by recurrent priapism [1]. Episodes are short lived (<4 hours), mainly occur at night, and are self-resolving. It is mainly seen in the homozygous sickle cell population and may affect up to 42% of men with sickle cell disease (SCD) [2–4].

Whereas there is broad agreement regarding the pathophysiology of classical low- or high-flow priapism, the mechanism of SP appears to be more

complex. Classical low-flow priapism in SCD is believed to be secondary to erythrocyte sickling, leading to venous and sinusoidal stasis in a closed compartment [5,6]. If a similar mechanism led to SP, then it fails to explain how an episode may self-terminate, as most episodes of SP do. Pathophysiological studies suggest that increased bioavailability of cyclic guanosine monophosphate (cGMP) levels in the corpus cavernosum, with enhanced smooth muscle relaxation in the corporal sinusoids, as a possible cause of SP [7]. Alteration of the normal penile hemodynamic equilibrium would better explain the transitory nature of SP, and rather than being a part of the low-flow/ischemic spectrum of priapism, SP may be a form of short-lived, high-flow priapism. Physiologically, this implies that the baseline blood flow may also be altered in this group. Imaging modalities such as penile Doppler ultrasound (PDU) may help evaluate the blood flow pattern in these patients. The hypothesis that resting cavernosal artery blood flow is altered in men with SCD and SP was studied using PDU.

Materials and Methods

Retrospective cohort study was carried out in a hospital with a specialist erectile dysfunction (ED) and andrology service. Patients who had undergone a PDU examination between April 2008 and April 2011 were identified by electronic interrogation of the radiological database. Our hospital does not require formal institutional review board approval or informed consent from patients for this type of study, as this was a retrospective study in which existing clinical data were used with no change in patient care (http://www.hra-decisiontools.org.uk/research/result7.html).

A standard study protocol was followed. Initial gray scale ultrasound examination was undertaken to evaluate the corpora cavernosa for fibrosis or calcification. Next, the baseline cavernosal artery flow was measured. The cavernosal artery enters the corpus cavernosum through the crus and for a short length (approximately 1–2 cm), the artery travels vertically through the proximal fixed part of the corpus cavernosum, before it curves into the pendulous portion of the corpus cavernosum. If a Doppler ultrasound probe is transversally placed at the junction of the fixed and pendulous portions, i.e., at the root of the flaccid penis, and tilted slightly cephalad, this vertical portion of the cavernosal artery can be readily seen (as shown in Figure 1). Baseline cavernosal artery flow is most easily measured in this vertical portion of the cavernosal artery. Furthermore, as arterial flow is directed vertically toward the probe, velocities can be measured at the most optimal Doppler angle of near zero. In each patient, the Doppler waveform was recorded and the baseline peak systolic and end-diastolic velocities were measured. Each Doppler waveform is a continuous temporal profile of the cavernosal artery velocity over a single cardiac cycle. The shape of the waveform and velocities reflect the vascular tone or peripheral resistance (PR) of the tissue bed supplied and can be categorized as high or low PR in shape (as further discussed below).

Those patients with ED or Peyronie's disease also underwent a poststimulation study after intracavernosal injection of prostaglandin E1 (median dose 10 μg). Because of the risk of priapism, men with a history of SP did not undergo pharmacostimulation, except one patient who was given only 1 μg of prostaglandin. For the study hypothesis, only the unstimulated PDU findings were compared. Clinical data were retrieved from the case notes. Where applicable, the diagnosis of SCD was confirmed by hemoglobin electrophoresis in all relevant cases. Data gaps were resolved by telephone interview of patients with SP. For comparison purposes, baseline cavernosal artery velocity was measured in 20 men with normal erectile function. These men were undergoing scrotal ultrasound for unrelated reasons, and resting cavernosal artery velocity was measured at the end of the scrotal examination after verbal consent from each case. All 20 men reported no erectile difficulty on direct questioning and more detailed inquiry was not conducted. Only resting cavernosal artery velocity was measured and pharmacostimulation was not given.

Results

Over the study period, 100 men had undergone PDU; 4 had Peyronie's disease (age range 35–48 years), 9 had SP (age range 20–40 years), 20 men had normal erections (age range 18–42 years), and the rest had ED (age range 16–67 years). There was no significant difference in the age distribution of these groups ($P \geq 0.05$; unpaired t-test). Twenty-five patients had homozygous SCD and the nine men with SP also had SCD. The other 16 men with SCD presented with ED, and all had given a prior history of priapism.

Men with SP were aged 20–40 years, of Afro-Caribbean descent, and reported that SP occurred