JAMA Clinical Guidelines Synopsis

Diagnosis and Management of Priapism

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GUIDELINE TITLES Priapism in People With and Without Sickle Cell Disease: Acute Diagnosis and Treatment

RELEASE DATES November 2021 and July 2022

DEVELOPER AND FUNDING SOURCE American Urological Association (AUA) and Sexual Medicine Society of North America (SMSNA)

TARGET POPULATION Males, including those with hematologic and oncologic disorders (eg, sickle cell disease [SCD]; chronic myelogenous leukemia [CML]) and those using intracavernosal vasoactive medications

MAJOR RECOMMENDATIONS

- Clinicians should counsel patients with a priapism duration >36 hours that recovery of erectile function is unlikely (moderate recommendation; evidence level B).
- Diagnostic testing should be done to determine the etiology of acute ischemic priapism (IP), but should not delay definitive treatment (expert opinion).
- Penile corporal blood gas should be measured at presentation to distinguish IP from nonischemic priapism (NIP) (expert opinion).
- First-line therapy for acute IP should be intracavernosal phenylephrine and corporal aspiration, with or without irrigation, before operative interventions (moderate recommendation; evidence level C).
- A distal corporoglanular shunt procedure should be performed if acute IP persists after intracavernosal phenylephrine and corporal aspiration, with or without irrigation (moderate recommendation; evidence level C).
- In patients with hematologic and oncologic disorders (eg, SCD, CML), standard management of acute IP should not be delayed for disease-specific systemic interventions (ie, exchange transfusion) (expert opinion).

Summary of the Clinical Problem

Priapism (an erection lasting >4 hours) results in 5.3 emergency department visits per 100 000 patient-years in the US.[1,2] Acute ischemic priapism (IP) is an emergent condition requiring urgent intervention within 6 to 12 hours of onset to prevent permanent erectile dysfunction, penile fibrosis, and penile shortening.[3] Up to 42% of men with SCD experience priapism during their lifetime.[4] This guideline describes approaches to diagnosis and management of IP and NIP, including in men with hematologic and oncologic disorders.

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Characteristics of the Guideline Source

This joint guideline was created by the AUA and SMSNA with funding from the AUA (**Table**). The panel was composed of urologists with topical expertise with representation from the American College of Emergency Physicians. Panel members disclosed potential conflicts of interest. Prespecified AUA nomenclature was used to formulate recommendation statements based on level of certainty, magnitude of benefit or risk/burden, and literature strength of evidence.

Evidence Base

Evidence-based practice center teams provided an evidence base on acute and recurrent IP and NIP.[2,3] Patient history should include baseline erectile function, degree of pain, history of priapism and treatment, use of drugs associated with priapism (eg, vasoactive erectile agents, antidepressants, α-blockers, trazodone, and illicit drugs), history of pelvic or genital trauma, family history of SCD or other blood abnormalities, and any history of malignancy. Erections associated with IP are typically persistently painful, while those associated with NIP are nearly always painless. The physical examination should establish whether the corpora cavernosa are fully rigid with sparing of the corpora spongiosum and glans penis (suggesting IP) or less than fully rigid (more typical of NIP). In most circumstances, corpus cavernosal blood gas should be measured on initial presentation using a butterfly needle of no smaller than 21 gauge.[6] This test does not require anesthesia and helps distinguish acute IP (typical values: Po_2 <30 mm Hg; Pco_2 >60 mm Hg; and pH <7.25) from NIP (typical values: Po_2 >90 mm Hg; Pco_2 <40 mm Hg; pH = 7.4). Additional laboratory testing may include a complete blood cell count along with blood and urine screening for psychoactive medications and illicit drugs. In acute evaluation and management of priapism by nonurologists in the emergency department, imaging studies are not recommended.

When acute IP is diagnosed, the clinician should begin rapid first-line treatment with corporal aspiration and intracavernosal phenylephrine (off-label use), which can be complemented by corporal irrigation if the clinician chooses. These procedures are performed after administering local anesthetic in the form of a penile or ring block. One retrospective review reported that successful detumescence occurred significantly more often with intracavernosal phenylephrine as initial treatment vs oral or subcutaneous terbutaline (17/23 [74%] vs

Table. Guideline Rating[5]

Standard	Rating
Establishing transparency	Good
Management of conflict of interest in the guideline development group	Good
Guideline development group composition	Fair
Clinical practice guideline-systematic review intersection	Good
Establishing evidence foundations and rating strength for each of the guideline recommendations	Good
Articulation of recommendations	Fair
External review	Fair
Updating	Fair
Implementation issues	Fair

2/8 [25%]; risk ratio, 0.34; 95% CI, 0.099-1.15; $P = .03$).[7] For patients with IP who do not improve with intracavernosal phenylephrine and aspiration, with or without irrigation, a distal corporoglanular shunt procedure should be performed. Shunts create an exit for blood trapped in the corpora cavernosa and help reestablish arterial penile blood flow. Shunts are created by surgically opening the distal end of the corpora cavernosa using a needle or scalpel. Some patients require concurrent corporal tunneling, in which an instrument is passed into the corporal tissue to facilitate better penile decompression.

Standard corpora cavernosa management of acute IP in patients with known or suspected hematologic and oncologic disorders should not be delayed while arranging systemic treatments such as exchange transfusions for the underlying disease, since these therapies are often futile in the treatment of IP.

In contrast to IP, NIP is characterized by high-flow arterial blood, and the penile tissue is not at risk of ischemic damage. Clinicians should advise patients that NIP is not an emergency condition and offer patients an initial period of observation with cessation of associated medications, unless a patient is severely bothered by the tumesced penis. In the absence of rigorous data on the optimal duration of observation, a 4-week monitoring period is suggested and will permit trending of a patient's sexual function and tolerance of priapism, after which interventional radiology embolization can facilitate resolution.

Benefits and Harms

Detumescence rates are as high as 93% with rapid first-line therapy of intracavernosal phenylephrine with aspiration (with or without irrigation), and 70% to 92% of patients report preserved erectile function afterward. The procedure can cause hypertension and reflex bradycardia; therefore, blood pressure and heart rate should be monitored. Case reports of myocardial infarction after intracavernosal phenylephrine administration have been published.[2,3,8]

Distal corpus cavernosal surgical shunt creation, with or without tunneling, is reserved for patients who do not improve with first-line treatments or for more than 48 hours after onset of priapism. These procedures can salvage unassisted erections in 15% of IP cases and erections responsive to oral medications in another 31% of men (depending on priapism duration and type of shunt).[9] Without distal corpus cavernosal surgical shunt creation, virtually all patients with persistent IP will have complete erectile dysfunction. Additionally, shunting can lead to complications such as infection, skin necrosis, urethral injury, fistula formation, and cavernositis.[2,3] A recent systematic review provided additional data on surgical and minimally invasive treatment of IP and NIP.[10]

Discussion

This guideline emphasizes the consequences of delaying treatment for acute IP, while providing a framework for managing this challenging condition. Given the numerous, diverse causes of priapism outlined in the guideline appendix, the challenges in treating priapism often extend beyond the pathology itself.[2,3] This is especially the case with recurrent priapism, which disproportionately affects patients at risk of marginalization from the health care system, such as those with lower income, with SCD, or who use illicit drugs.[2]

After the patient's history and physical examination are completed, baseline corporal blood gas should be tested to delineate acute IP from NIP.[2,3] Conservative therapies such as exercise, cold compresses, and oral medications do not effectively treat IP and should not be used.[2,3] Intravenous fluid resuscitation, supplemental oxygen, or exchange transfusion should occur promptly when indicated but should not delay definitive therapy for IP.[2,3] If first-line methods are ineffective, prompt distal corporoglanular shunting should be done to prevent further tissue ischemia and necrosis.[2,3]

Areas in Need of Future Study or Ongoing Research

Additional basic, translational, and clinical outcomes research are needed to advance therapy in this field. Preventive strategies including use of the monoclonal antibody crizanlizumab for priapism in SCD are being explored for management of stuttering priapism, which is intermittent and recurring IP with episodes commonly lasting less than 4 hours. Improved approaches to support shunt patency after corporoglanular shunt procedures and mitigate thrombosis are needed. Based on lower-level evidence (expert opinion), the guideline suggests consideration of placing a penile prosthesis at the time of an acute IP episode lasting more than 36 hours or in those refractory to shunting. Finally, penile embolization techniques are commonly used to treat men with persistent NIP. Further studies are needed to refine embolization materials and techniques, addressing both short- and long-term outcomes.

ARTICLE INFORMATION

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REFERENCES

1. Roghmann F, Becker A, Sammon JD, et al. Incidence of priapism in emergency departments in the United States. *J Urol*. 2013;190(4):1275-1280. doi:10.1016/j.juro.2013.03.118

2. Bivalacqua TJ, Allen BK, Brock GB, et al. The diagnosis and management of recurrent ischemic priapism, priapism in sickle cell patients, and non-ischemic priapism. *J Urol*. 2022;208(1):43-52. doi:10.1097/JU.0000000000002767

3. Bivalacqua TJ, Allen BK, Brock G, et al. Acute ischemic priapism. *J Urol*. 2021;206(5):1114-1121. doi:10.1097/JU.0000000000002236

4. Nelesen D, Lucas S, Liu C-R, et al. A systematic review to assess the burden of ischemic priapism in patients with sickle cell disease. *Sex Med Rev*. 2023;11(1):52-60. doi:10.1093/sxmrev/qeac001

5. Cifu AS, Davis AM, Livingston EH. Introducing JAMA Clinical Guidelines Synopsis. *JAMA*. 2014;312(12):1208-1209. doi:10.1001/jama.2014.12712

6. Reed-Maldonado AB, Kim JS, Lue TF. Avoiding complications: surgery for ischemic priapism. *Transl Androl Urol*. 2017;6(4):657-665. doi:10.21037/tau.2017.07.23

7. Martin C, Cocchio C. Effect of phenylephrine and terbutaline on ischemic priapism. *Am J Emerg Med*. 2016;34(2):222-224. doi:10.1016/j.ajem.2015.10.029

8. Constantine ST, Gopalsami A, Helland G. Recurrent priapism gone wrong: ST-elevation myocardial infarction and cardiogenic shock after penile corporal phenylephrine irrigation. *J Emerg Med*. 2017;52(6):859-862. doi:10.1016/j.jemermed.2017.01.055

9. Ortaç M, Çevik G, Akdere H, et al. Anatomic and functional outcome following distal shunt and tunneling for treatment ischemic priapism. *J Sex Med*. 2019;16(8):1290-1296. doi:10.1016/j.jsxm.2019.05.011

10. Milenkovic U, Cocci A, Veeratterapillay R, et al. Surgical and minimally invasive treatment of ischaemic and non-ischaemic priapism. *Int J Impot Res*. Published online September 23, 2022. doi:10.1038/s41443-022-00604-1