

A Whittington Hospital Clinical Management Guideline

## PRIAPISM IN SICKLE CELL ANAEMIA

**Date:** October 2006  
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**Directorate:** Medicine  
**Owner:** Drs Rai/ Bielawski

**Relevant to:** All medical and nursing staff

**Key words:** Sickle cell anaemia, priapism, painful erection, penile aspiration

**Note:** It is imperative that the consultant haematologist on call is informed about all sickle cell patients who present with acute priapism.

### Contents of this Guidance:

- General management of acute priapism (**section I**)
- Specific management of acute priapism greater than 4 hours duration (**Section II**)
- Management of stuttering priapism (**section III**)
- Outpatient referral form for the urology unit (**Appendix 1**)
- Urgent transfer referral form for the urology unit (**Appendix2**)
- Patient information leaflet (**Appendix 3**)
- International Index of Erectile Function (IIEF) score sheet (**Appendix 4**)

### Criteria for use

The procedures outlined in this guideline must be followed by general medical, urology, haematology and emergency department staff for all male patients with sickle cell anaemia complaining of painful prolonged erection that does not resolve spontaneously.

## Background/ introduction

Priapism is a condition where penile erection persists beyond, or is unrelated to sexual stimulation and occurs in 40% of men with sickle cell anaemia. It is a type of compartment syndrome and therefore a surgical emergency. Priapism is very common and needs to be looked for and treated carefully. Failure to appropriately manage priapism or to recognise it can lead to impotence.

Stuttering priapism is a condition where the erection typically lasts for less than 4 hours and often will precede the development of an acute priapism.

Beyond four hours this is a surgical emergency and should be treated as an acute ischaemic priapism.

## SECTION ONE: General management

### THE FOLLOWING SPECIFIC POINTS MUST BE RECORDED IN ALL PATIENT NOTES

#### History

1. Time of arrival at hospital
2. Duration of priapism ( hrs: min)
3. Measure to relieve priapism at home:
  - Exercise
  - Masturbation/ sex
  - Other
4. Precipitating factors:
  - Trauma
  - Dehydration
  - Prescribed medication
  - Alcohol
  - Non-prescription drugs
5. IIEF score – which gives information about previous erectile function

#### Examination

1. Confirm priapism - Tender rigid penile shaft with soft glans
2. Bladder palpable - Encourage passing urine, catheterise if attempts fail
3. Bladder not palpable - Proceed to Specific management

**THE FOLLOWING STEPS SHOULD BE FOLLOWED REGARDLESS OF DURATION OF PRIAPISM.**

- Inform Urology team. Instigate the following measures prior to reply:
- Analgesia (as per patients individual sickle cell analgesia protocol)
- Oral hydration
- Exercise e.g. brisk walk, jogging on the spot
- Oral etilefrine\*: Adult 50 mg bd  
Paediatric 0.25mg/kg bd
  - continue oral etilefrine until patient reviewed by consultant urologist at UCH (Mr Minhas) or deputy ( Appendix 1)

*\* Pseudoephedrine should be used if etilefrine is not available ( adults 30-60mg qds; children 15-60 mg qds according to age as per BNF)*

**IF THESE MEASURES RELIEVE THE PRIAPISM ADMIT UNDER THE UROLOGY TEAM FOR OBSERVATION OVERNIGHT**

- Ensure regular prescription of oral etilefrine or pseudoephedrine as above
- Monitor BP
- Fax referral form to consultant urologist at UCH (Mr Minhas) or deputy (Appendix 1)
- Patient must be discharged with a copy of referral letter, an appointment for the above clinic and a patient information leaflet (Appendix 3)

**SECTION 2: Specific management of acute priapism greater than 4 hours duration**

**Urologist** to obtain cavernosal blood gas leaving butterfly in situ (*imperative blood gas performed on first sample drawn from penis*)

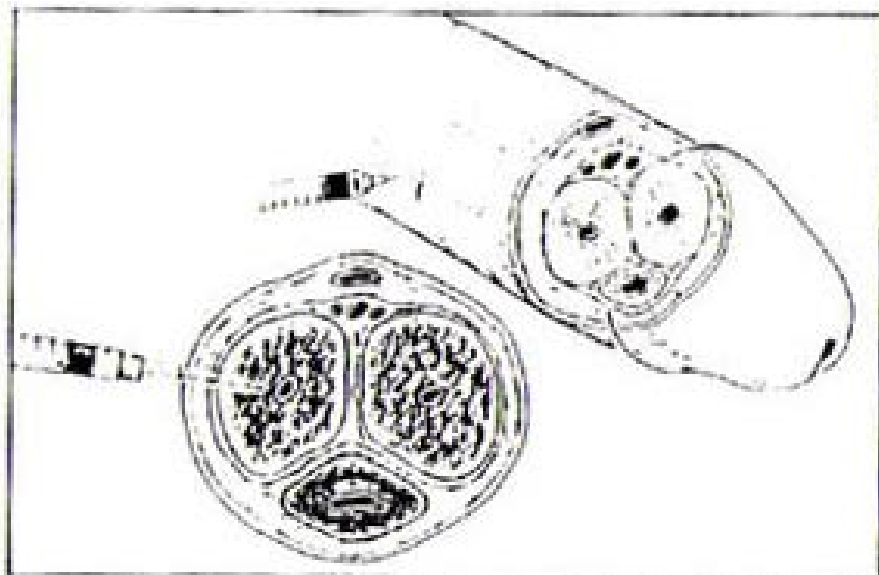
## INTRACAVERNOSAL BLOOD ASPIRATION AND INJECTION

You will need:

- Urethral catheterisation pack (if not available then dressing pack).
- Liquid skin antiseptic (chlorhexidine/povidone iodine)
- Green Butterfly needle (19 G needle)
- Heparinised (blood gas) syringe
- 20ml syringe for aspiration
- 10ml syringe for injection
- Phenylephrine 1mg made up to 10ml with normal saline/water for injection

### PROCEDURE:

1. Written consent for procedure. Risks include pain and bleeding
2. Lay patient flat
3. Clean penis with antiseptic liquid
4. Hold penis by glans and insert needle into side of penis at midshaft level to a depth of 1cm (see diagram). Care should be taken to avoid the urethra and neurovascular bundle.
5. Aspirate blood with heparinised syringe and process through blood gas machine. At same time, send separate sample of penile blood in fluoride tube to Biochemistry for measurement of glucose.
6. Aspirate **up to 100ml blood** from penis with 20ml syringe. Blood should be thick and dark. Once blood becomes fresh stop.
7. Inject phenylephrine as described below whilst monitoring the cardiovascular system.



## **IF PRIAPISM RELIEVED ADMIT OVERNIGHT FOR OBSERVATION**

- Ensure regular prescription of oral etilefrine as above
- Monitor BP
- Fax referral form to consultant urologist at UCH (Mr Minhas) or deputy (Appendix 1)
- Patient must be discharged with a copy of referral letter, an appointment for the above clinic and a patient information leaflet (Appendix 3)

**If the penis after injection fails to subside- the patient will require immediate Blue light**

**Contact on call-Urology SpR at UCLH and send urgent referral letter (appendix 2) with the patient.**

**Arrange Blue light to UCLH**

### **SECTION 3: Stuttering Priapism**

Follow section one on general management and start patient on prophylaxis as above.

Additional tests should be taken for FBC/LFTs/FSH/LH/Testosterone

**It is imperative that an urgent referral is faxed to consultant urologist at UCH (Mr Minhas) or deputy (Appendix 1)**

### **Contacts (inside and outside the Trust including out-of-hours contacts)**

Dr Norman Parker  
Consultant Haematologist  
Whittington Hospital  
0207 288 5437

Dr Bernard Davis  
Consultant Haematologist  
Whittington Hospital  
0207 288 5035

Dr Farrukh Shah  
Consultant haematologist  
Whittington Hospital  
0207 288 5144

Professor John Porter  
Consultant Haematologist  
UCLH  
0207 679 6224 day-time  
0845 1555 000

Mr S Minhas  
Consultant Urologist  
UCLH  
0845 1555 000

Blood Transfusion Lab  
Whittington Hospital  
(via switchboard: 0207 272 3070)

Urology SpR on call  
UCLH  
via mobile

Urology ward UCLH T7 South  
0845 1555 000  
extension 70700 or 70782

## References (evidence upon which the guideline is based)

1. Kumar et al. The surgical management of priapism: Curr Sex Health Rep 2004: 1: 125-128
2. Adeyoju et al. Priapism in sickle-cell disease, risk factors and complications-an international multicentre study: BJUI 2002: 90: 898-902
3. Pryor et al. Priapism (WHO guidelines): J Sex Med 2004: 1: 116-119
4. Montague et al. AUA guideline on the management of priapism: J Urol 2003: 170: 1318-1324
5. Burnett et al. Evaluation of erectile function in men with sickle cell disease: Urology 1995: 45: 657-663
6. Okpala et al. Etilefrine for the prevention of priapism in adult sickle cell disease: B J Haem 2002: 118: 918-921
7. Gbadoé et al. Management of sickle cell priapism with etilefrine: Arch Dis Child 2001: 85: 52-53
8. Rosen et al. The International index of erectile function (IIEF): A multidimensional scale for assessment of erectile dysfunction: Urology 1997: 49: 809-988

**Appendix 1: Outpatient referral form if priapism relieved or stuttering priapism**

**The Whittington Hospital** 

NHS Trust

*Highgate Hill  
London, N19 5NF*

*Tel. 020 7272 3070 - switchboard*

Today's date:

**FAX TO: 020 7380 9063**

Dear Mr Minhas

Name:

Address:

DOB:

Hospital Number:

Telephone number:

I would be grateful if you could see this patient urgently in your specialist clinic. The details are noted below pertaining to the priapic episode.

Time of arrival at hospital	
Duration of priapism ( hrs: min)	
Measures to relieve priapism at home 1. Exercise 2. Masturbation/ sex 3. Other	
Precipitating factors: 1. Trauma 2. Dehydration 3. Prescribed medication 4. Alcohol 5. Non prescription drugs	
Previous episodes last 6 months	
IIEF SCORE	

The penile blood gas was:

pO<sub>2</sub> :

pCO<sub>2</sub>

pH:

glucose:

The priapism was relieved after the following interventions:

- 1. Analgesia
- 2. Exercise
- 3. Hydration
- 4. Etilerfrine/Pseudoephedrine
- 5. Penile aspiration
- 6. Penile injection of phenylephrine

The patient is now taking oral prophylaxis in the form

of:.....  
.....

Please can you arrange for him to be reviewed in your clinic.

Yours sincerely

Name:

Post:

Contact number:

*Copy to Dr F Shah  
Haematology Consultant  
Whittington Hospital  
Highgate Hill  
London  
N19 5NF*



## URGENT Sickle Priapism

Today's date:

**FAX TO: 020 7380 9063**

Dear Urology SpR

RE:

Name:

Address:

DOB:

Hospital Number:

Telephone number:

Has presented to Whittington hospital with an acute priapism that has failed to resolve. Please see the details of management so far:

Time of arrival at hospital	
Duration of priapism ( hrs: min)	
Measures to relieve priapism at home 4. Exercise 5. Masturbation/ sex 6. Other	
Precipitating factors: 6. Trauma 7. Dehydration 8. Prescribed medication 9. Alcohol 10. Non prescription drugs	
Previous episodes last 6 months	
IIEF SCORE	

The penile blood gas was:

pO<sub>2</sub> :

pCO<sub>2</sub>

pH:

glucose:

The priapism was not relieved despite:

- 1. Analgesia
- 2. Exercise
- 3. Hydration
- 4. Etilefrine/Pseudoephedrine
- 5. Penile aspiration
- 6. Penile injection of phenylephrine

Details of penile aspiration

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Attached is patient specific analgesia protocol from Whittington Hospital:

Yours sincerely

Name:

Post:

Contact number:

*Copy to Dr F Shah  
Haematology Consultant  
Whittington Hospital  
Highgate Hill  
London  
N19 5NF*

### Appendix 3: Patient information leaflet

#### **Painful erections in men with sickle cell anaemia, also known as priapism.**

What is it?

How do I recognise the problem?

What can I do about it?

This leaflet aims to give you some of the answers.

Most boys over the age of about 10 years and healthy adults will have at least 3 – 4 erections a day. Many will happen at night and it is normal for older boys and men to wake up with an erection. Normal erections are not painful and fade away as the person gets on with their normal life. If the erection is painful or if it does not fade away after about 30 minutes this is called priapism. Priapism is very common in boys and men with sickle cell and needs to be treated carefully. If the problem is ignored it may lead to future problems; at worst the man may not be able to have normal erections.

Do the following things immediately

- Drink lots of fluids
- Some patients find jogging or other exercise helps
- Take simple pain killers such as paracetamol
- Try to ejaculate

This may take the problem away within the next hour, if so you should arrange to report the problem to the team caring for you within the next few days.

If the problem does not go away you should go to the emergency department. Show them this leaflet and ask them to follow the instructions that have been left there for dealing with the problem. The plan will include a series of steps.

In almost all cases a sample of blood will be taken from the penis to check how much acid and oxygen is in the blood.

Other steps may include:

- A medicine – etilefrine by mouth or perhaps an injection into the penis.
- Draining some blood from the penis.
- A different drug which will stop erections by lowering testosterone is used in some cases
- In severe cases you may need an operation done by a specialist team at University College Hospital. There are several types of operation and these will be discussed with you.

## Appendix 4: International Index of Erectile Function (IIEF) Score

Table showing questions required to obtain IIEF score<sup>8</sup>  
 Add score from each question to obtain final total

1. How do you rate your <u>confidence</u> that you could get and keep an erection?					
Very low 1	Low 2	Moderate 3	High 4	Very high 5	
2. When you had erections with sexual stimulation, <u>how often</u> were your erections hard enough for penetration (entering your partner)?					
No sexual activity  0	Almost never or never  1	A few times (much less than half the time)  2	Sometimes (about half the time)  3	Most times (much more than half the time)  4	Almost always or always  5
3. During sexual intercourse, <u>how often</u> were you able to maintain your erection after you had penetrated (entered) your partner?					
Did not attempt intercourse  0	Almost never or never  1	A few times (much less than half the time)  2	Sometimes (about half the time)  3	Most times (much more than half the time)  4	Almost always or always  5
4. During sexual intercourse, <u>how difficult</u> was it to maintain your erection to completion of intercourse?					
Did not attempt intercourse  0	Extremely difficult  1	Very difficult  2	Difficult   3	Slightly difficult  4	Not difficult  5
5. When you attempted sexual intercourse, <u>how often</u> was it satisfactory for you?					
Did not attempt intercourse  0	Almost never or never  1	A few times (much less than half the time)  2	Sometimes (about half the time)  3	Most times (much more than half the time)  4	Almost always or always  5