Whittington Health NHS

Priapism in sickle cell anaemia, Management of

Version:	1.3
Ratified by:	Clinical Guidelines Committee
Date ratified:	July 2006, minor change September 2011, reviewed October 2014 with no change. Reviewed 2019 updated doses
Name of originator/author:	Dr Farrukh Shah
Name of responsible committee/individual:	As above
Date re-issued:	February 2019
Review date:	3 years hence
Target audience:	All junior doctors and consultants medical and surgical, ED staff

Revision Chronology:		
Version Number	**Effective date**	**Reason for Change**
1.1	**September 2011**	**Change in phone numbers for UCLH**
1.2	**October 2014**	**Reviewed according to date. No change required (Dr F Shah)**
1.3	**Feb 2019**	**Evidence base updated**

		Change of dose of etilefrine and psuedoephredine **Change of dose and reconstitution of phenylephrine**

A Whittington Health Clinical Management Guideline

Priapism in sickle cell anaemia, Management of

Relevant to: *All staff groups*

Key words: Sickle cell anaemia, Priapism, painful erection, penile aspiration

➢ contents

➢ Criteria for use

The procedures outlined in this guideline must be followed 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 occurring 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.

➢ Objectives

This document aims to provide guidance for the safe and effective management of priapism in the UCLH setting and to encompass the quality standards for sickle cell (WMQRS, 2012).
This guideline applies to all staff involved in the management of people with priapism at WH.

➢ References and evidence base

- Standards for the Clinical Care of Adults with Sickle Cell Disease (Society, 2008a)and 2018
- Sickle Cell Disease in Childhood: Standards and Guidelines for Clinical Care (Programmes, 2010)
- RCN Competencies: Caring for people with sickle cell disease and thalassaemia syndromes - A framework for nursing staff (Nursing, 2011)
- Quality Standards: Health Services Caring for Adults with Haemoglobin Disorders (WMQRS, 2012&2014)
- Kumar et al. The surgical management of priapism: Curr Sex Health Rep 2004: 1: 125-128
- Priapism in sickle-cell disease, risk factors and complications- an international multicentre study (Adeyoju et al., 2002)
- Priapism (WHO guidelines) (Pryor et al., 2004)
- American Urological Association guideline on the management of priapism. (Montague et al., 2003)
- Evaluation of erectile function in men with SCD. (Burnett et al., 1995)
- Etilefrine for the prevention of priapism in adult sickle cell disease. (Okpala et al., 2002)
- Management of sickle cell priapism with etilefrine. (Gbadoe et al., 2001)
- The International index of erectile function (IIEF): A multidimensional scale for assessment of erectile dysfunction. (Rosen et al., 1997)

SECTION ONE: general management

THE FOLLOWING SPECIFIC POINTS MUST BE RECORDED IN ALL PATIENTS NOTES (Medico-legal implications)

History

Time of arrival at hospital	
Duration of priapism (hrs: min)	
Measure 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	
Prior IIEF SCORE (Appendix 4)	

Examination

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

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

Inform Urology team; do not wait for consult to instigate following measures

PHYSICAL:

- Analgesia (as per patients individual sickle cell analgesia protocol)
- Oral hydration
- Exercise e.g. brisk walk, jogging on the spot
- Bladder- if in doubt perform bladder scan on ward

If palpable - Encourage passing urine, catheterise if attempts fail

MEDICATION:

- Oral etilefrine*: Adult 50 mg TDS
  - Paediatric 0.25mg/kg po in 3 divided doses

- Psuedoephidrine should be used if etilefrine is not available (30-60mg qds)
  - Peadiatric 15-60 mg QDS according to age (BNF)

IF THESE MEASURE RELEIVE THE PRIAPISM ADMIT UNDER THE UROLOGY TEAM FOR OBSERVATION OVERNIGHT

- Ensure regular prescription of oral etilefrine or pseudoephedrine as above
- Monitor BP and manage in accordance with Trust Guidelines for Management of Sickle Cell Disease in Adulthood: General Management and Childhood
- Patient must be discharged with
  - An outpatient appointment under the Andrologists at UCLH
  - A patient information leaflet

	Please see Whittington Hospital NHS Trust Guideline: ***Sickle Cell Disease in Adults***

	Please see Whittington Hospital NHS Trust Guideline: ***Sickle Cell Disease in Childhood***

	Please see Whittington Hospital NHS Trust Patient information leaflet: ***Priapism for patients with Sickle Cell Disease*** http://whittnet.whittington.nhs.uk/document.ashx?id=2272

IF THESE MEASURES **DO NOT** RELIEVE THE PRIAPISM ADMIT UNDER THE UROLOGY TEAM AND FOLLOW ADVICE ACCORDING TO SECTION 2

UROLOGICAL EMERGENCY REQUIRING HAEMATOLOGY SUPPORT

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

HAEMATOLOGY:

- Make arrangements for a potential exchange blood transfusion (see Trust Guidelines for Management of Sickle Cell Disease in Adulthood: General Management and Childhood
- **Inform Haematology Consultant on Call**

Urological treatment of the priapism which should not be delayed

UROLOGY:

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)
- 2x White Butterfly needles (19 G needle)
- Heparinised (blood gas) syringe
- 2 x 60mls syringes for aspiration
- 20ml syringe for injection
- Phenylephrine 1 vial(10mg) in 19mls saline, 0.5 mls at a time
- 1% lidocine x 10mls
- 10ml syringe and 23g needle
- An assistant
- Consider general anaesthetic for small children

PROCEDURE:

1 Written consent for procedure. Risks include pain and bleeding

2 Lay patient flat

3 Clean penis with antiseptic liquid

4 Inject bleb of lidocaine at proposed aspiration site on both sides of the shaft

5 Hold penis by glans and insert butterfly needles into each side of penis at midshaft level to a depth of 1cm (see diagram). Care should be taken to avoid the urethra and neurovascular bundle.

6 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.

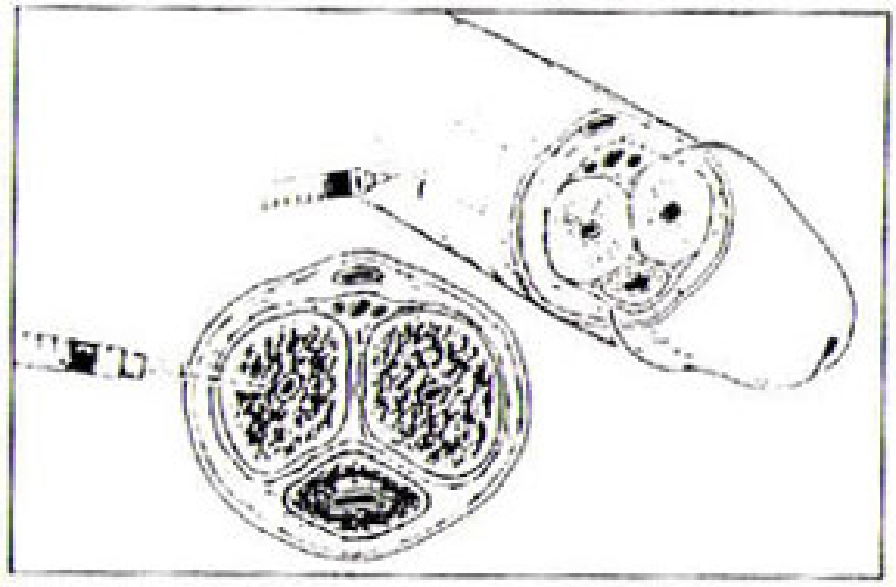
7 Aspirate up to 100ml blood from penis with 60ml syringe. Blood should be thick and dark. Once blood becomes fresh, stop. Wash with heparin saline through the 2nd butterfly needle until all dark blood aspirated

8 If detumescence has occurred just observe

9 If penis becomes erect again proceed to phenylephrine injection through the butterfly needle

10 Inject phenylephrine in 0.5ml aliquots repeated every 3 minutes for 1 hour whilst monitoring the pulse and blood pressure

11 Compress penis after needle removal for 2 minutes



IF PRIAPISM RELIEVED ADMIT OVERNIGHT FOR OBSERVATION

- Ensure regular prescription of oral etilefrine or Psuedoephidrine as above
- Monitor BP
- Arrange outpatient follow-up with Consultant Andrologists
- Patient must be discharged with an appointment for the Andrology clinic at UCLH and a patient information leaflet

PERSISTENT PRIAPISM
IF THE PRIAPISM, AFTER INJECTION, FAILS TO SUBSIDE THEN SURGICAL PROCEDURE IS USUALLY NEEDED.

- This will often necessitate an exchange blood transfusion
- Current management is a T shunt if within 48 hours
- If longer than 3 days then an MRI of the penis should be performed to assess smooth muscle perfusion
- Consider penile prosthesis T insertion for refractory priapism or fibrosis from previous events.

ANTIBIOTICS
Please refer to the Trust Urology infections – treatment and prophylaxis guideline for guidance on choice of antibiotic, dose and duration of therapy.

Contact on call-Urology SpR at UCLH and over relevant details from appendix 2.

Arrange Blue light to UCLH

SECTION 3: Stuttering Priapism

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

- Andrology review and decision whether management according to Section 2 is appropriate.
- Optimise haematological status
- Commence on regular etilefrine /pseudoephedrine
- Ensure patient has urgent referral and appointment with Andrology team at UCLH. They should present to ED at UCLH if they develop a Priapism which is not resolving in the time between review at Whittington and Appointment at Andrology clinic.

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

SECTION 4: blood transfusion

Blood Transfusion

- There is no randomised trial for administering an exchange blood transfusion in the case of priapism in sickle cell disease. Please

see Trust Guidelines: Management of Sickle Cell Disease in Adulthood: Specific Treatments.

- However, it is often deemed an appropriate treatment in difficult cases and may well be relevant in any case if the patient is to go to theatre.
- Exchange transfusions require excellent venous access and significant volumes of blood. They also require substantial staff time.
- This is why it is imperative that the Red Cell haematology team is involved from the outset i.e. from when they attend in A+E or in the case of tertiary referrals from when the andrology team is called from the referring centre. Blood and central venous access can be organised prior to the patient coming to the hospital
- The decision to exchange will be made by the haematology team in conjunction with one of the consultants, though the Andrology team may want to recommend it.

SECTION 5: contact details for Whittington services

Haematology consultants via Switchboard

Haematology Spr: bleep 3060 and 3036

SECTION 6: contact details for UCLH services

Red Cell Haematology SHO: 7042, Out of hours: 7003

Red Cell Haematology SpR: 7085, Out of hours: hospital switchboard

Red Cell Consultant: switchboard

Andrology: Andrology SpR and Consultant through switchboard (Mr Minhas or Mr Ralph are happy to be contacted directly – email addresses on outlook or through switchboard)

Joint Clinic: there is a joint haematology/andrology clinic running on Wednesday evenings in the Cancer centre DJR3P (Mr Ralph) and JBP3E (Professor Porter

Appendix 1: Outpatient referral Protocol if priapism relieved or stuttering priapism

Consultants

Professor J B Porter MA MD FRCP FRCPath
Dr Sara Trompeter BSc MRCPCH FRCPath
Dr Emma Drasar FRCPath MRCP PhD
Dr Bernard Davis MD FRCP, FRCPath
Dr Farrukh Shah MD FRCP,FRCPath
Dr Perla Eleftheriou FRCP, FRCPath
Dr Ratna Chatterjee MD PhD MFFP
Dr Andrew Robins MSc, MRCP, FRCPCH
Nursing team:
Bernadette Hylton
Nancy Huntlry
Emma Prescott
Edith Aimiuwu
Matty Asante-Owusu

Whittington Health NHS

RED CELL DISORDERS UNIT
Jointly with UCLH
Department of Haematology

Magdala Avenue,London,N19 5NF
Switchboard: 020 7272 3070
Outpatient Access Centre 020 72885 5511

Mr Ralph
Consultant Andrologists
UCLH
250 Euston Road
London NW1 2PG
Date

Dear Mr Ralph /Minhas

Re:

Problems (list):

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:

pO2 : pH

pCO2 glucose:

The priapism was relieved after the following interventions:

1 Analgesia

2 Exercise

3 Hydration

4 Etilefrine/Pseudoephedrine

5 Penile aspiration

6 Penile injection of phenylephrine

The patient is now taking oral prophylaxis in the form

of:……..

Thank you for your help.

Yours sincerely

Name:

Post:

Copy to Dr E Drasar / Dr F shah, Dr B Davis

Consultant Haematologists Whittington hospital

Appendix 2: ACTE PRIAPISM URGENT TRANSFER REFERAL

Whittington Health NHS

Department of Haematology
Magdala Avenue,London,N19 5NF
Switchboard: 020 7272 3070
Outpatient Access Centre 020 72885 5511

URGENT
Sickle Priapism

Today's date:

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)	
Measure 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	

Prior IIEF SCORE (Appendix 3)	

The penile blood gas was:
pO_2 :
pCO_2
pH:
glucose:

The priapism was not relieved despite:

1. Analgesia ☐
2. Exercise ☐
3. Hydration ☐
4. etilefrine ☐
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/ Dr E Drasar or Dr B Davis
Haematology Consultant
Whittington Hospital
Magdala Avenue
London
N19 5NF

Appendix 3: Patient information leaflet

Please see Whittington Hospital NHS Trust
Patient information leaflet:
Priapism for patients with Sickle Cell Disease
http://whittnet.whittington.nhs.uk/document.ashx?id=2272

Appendix 4: IIEF Score

1. How do you rate your confidence that you could get and keep an erection?

Very low	Low	Moderate	High	Very high
1	2	3	4	5

2. When you had erections with sexual stimulation, how often were your erections hard enough for penetration (entering your partner)?

No sexual activity	Almost never or never	A few times (much less than half the time)	Sometimes (about half the time)	Most times (much more than half the time)	Almost always or always
0	1	2	3	4	5

3. During sexual intercourse, how often were you able to maintain your erection after you had penetrated (entered) your partner?

Did not attempt intercourse	Almost never or never	A few times (much less than half the time)	Sometimes (about half the time)	Most times (much more than half the time)	Almost always or always
0	1	2	3	4	5

4. During sexual intercourse, how difficult was it to maintain your erection to completion of intercourse?

Did not attempt intercourse	Extremely difficult	Very difficult	Difficult	Slightly difficult	Not difficult
0	1	2	3	4	5

5. When you attempted sexual intercourse, how often was it satisfactory for you?

Did not attempt intercourse	Almost never or never	A few times (much less than half the time)	Sometimes (about half the time)	Most times (much more than half the time)	Almost always or always
0	1	2	3	4	5

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Appendix A

Plan for Dissemination and implementation plan of new Procedural Documents

To be completed and attached to any document which guides practice when submitted to the appropriate committee for consideration and approval.

Acknowledgement: University Hospitals of Leicester NHS Trust

Title of document:	**Priapism in sickle cell anaemia**		
Date finalised:	**2006, re-issued sept 2011,October 2014,** **February 2019**	**Dissemination lead: Print name and contact details**	**Dr Shah**
Previous document already being used?	**Yes / No** **(Please delete as appropriate)**		
If yes, in what format and where?	**Electronic**		
Proposed action to retrieve out-of-date copies of the document:	**Removal from intranet site**		
To be disseminated to:	**How will it be disseminated/implemented, who will do it and when?**	**Paper or Electronic**	**Comments**
Urology Sprs	**Electronic**		
Urology consultants	**Electronic**		
Junior medical staff	**Electronic**		
Haematology team	**Electronic**		
Is a training programme required?	**no**		
Who is responsible for the training programme?			

Appendix B

Equality Impact Assessment Tool

To be completed and attached to any procedural document when submitted to the appropriate committee for consideration and approval.

Impact (= relevance) 1 Low 2 Medium 3 High	Evidence for impact assessment (monitoring, statistics, consultation, research, etc	Evidential gaps (what info do you need but don't have)	Action to take to fill evidential gap	Other issues
Race	**1**			
Disability	**1**			
Gender	**1**			
Age	**1**			
Sexual Orientation	**1**			
Religion and belief	**1**			

Once the initial screening has been completed, a full assessment is only required if:

- The impact is potentially discriminatory under equality or anti-discrimination legislation
- Any of the key equality groups are identified as being potentially disadvantaged or negatively impacted by the policy or service
- The impact is assessed to be of high significance.

If you have identified a potential discriminatory impact of this procedural document, please refer it to relevant Head of Department, together with any suggestions as to the action required to avoid/reduce this impact.