

Epidemiology
DEMO Priapism

Date Form Completed: **COMPDA** / **COMPMO** / **COMPYR**
COMPDT Day Month Year

Identifier acoustic: _____
(for internal use only)
Form completed by:

COMPLT

1. Patient's Date of Birth: **DOBDA** / **DOBMO** / **DOBYR**
DOBDT Day Month Year

2. Diagnosis: SS SC Sβ⁺thal Sβ⁰thal

3. Is this subject enrolled in C-Data (the CSCC collaborative database)? Yes No (If yes, skip Questions 4-6 and continue with Question 7.)

4. Date first seen in this center: / or Unknown
VISITMO Month **VISITYR** Year

5. Do you think that the medical history on this case report form is incomplete due to admissions at other hospitals that are not associated with your Center? Yes No Unknown

If yes, # years at other hospital(s)? **HOSPYR** List hospital(s), if known: **HOSPNAME**

6. From clinical records please indicate if, in his **entire** lifetime, this patient ever had or has been diagnosed with:

Splenic sequestrator	SPLSQ	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	SPLSQYR
Splenectomy?	SPTMY	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	SPTMYR
Dactylitis?	DACTL	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	DACTLYR
Leg ulcers?	ULCER	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	ULCERYR
Stroke?	AVASC	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	STRKEYR
Acute renal failure?	STRKE	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If known, year of 1 st Dx:	<input type="text"/> <input type="text"/>	ACUTEYR
Avascular necrosis?	ACUTE	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown			

If yes, list bone(s) involved and year(s) of first diagnosis (if known):

Bone: **BONE1** year of 1st Dx: **BONE1YR**

Bone: **BONE2** year of 1st Dx: **BONE2YR**

Bone: **BONE3** year of 1st Dx: **BONE3YR**

7. From clinical records please indicate if, in his **entire** lifetime, this patient ever had or has been diagnosed with:

Pulmonary hypertension? Yes No Unknown

If yes, list all known tests performed for diagnosis, procedures performed in relation to this condition, and year(s) of occurrence: **PULMTST**

Sickle cell retinopathy? Yes No Unknown

If yes, list all known tests performed for diagnosis, procedures performed in relation to this condition, and year(s) of occurrence: **SCELTST**

Chronic renal failure	CHRNC	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If yes, dialysis?	<input type="checkbox"/> Yes <input type="checkbox"/> N	DIALSIS
Admission for painful	PNCRS	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If yes, # times:	<input type="checkbox"/> <10 <input type="checkbox"/>	NUMPCRS
Acute chest syndrom	CHEST	<input type="checkbox"/> Yes	<input type="checkbox"/> No	<input type="checkbox"/> Unknown	If yes, # episodes:	___ In the last ___ 3 or more	ACSLST3 ACS3AGO

8. Most recent steady state lab results and date tests performed:

Hemoglobin: HMGLBN g/dL

HGBDA / HGBMO / HGBYR **HGBDT**
 day month year

Platelet count: PLTLET x10⁹ /L

PLTDA / PLTMO / PLTYR **PLTDT**
 day month year

WBC: WBC x10⁹ /L

WBCDA / WBCMO / WBCYR **WBCDT**
 day month year

Room air O₂ Sat: O2SAT % **OR** Not done/ Data not available

O2SATND O2SATDA / O2SATMO / O2SATYR **O2SATDT**
 day month year

Method used for determination: Pulse oximeter Other, specify: O2METHS
O2METH

9. Has the patient ever been placed on chronic transfusion therapy (planned transfusion about once per month for 3 months or more)? Yes No Unknown **CTRAN**

9a. If yes, what happened to the frequency of priapism during chronic transfusion therapy? **CTRANYS**

Increased Decreased Stayed about the same Don't know Patient was not having episodes of priapism when chronic transfusion was started

10. Has this patient ever taken Hydroxyurea? Yes No Unknown **HYDRX**

10a. If yes, what happened to the frequency of priapism during Hydroxyurea therapy? **HYDRXYS**

Increased Decreased Stayed about the same Don't know Patient was not having episodes of priapism when Hydroxyurea was started

11. Was your clinical program aware, by a mention in the medical record, that this patient had ever had priapism before this questionnaire was conducted? Yes No **AWARE**

11a. If yes, how has it been managed or treated? (check all that apply)

NARCTC Narcotics and hydration for episodes **TRNSFSN** Transfusion (simple or chronic) **ASPIRRG** Aspiration and irrigation for prolonged episodes **PSEUDO** Pseudoephedrine for prevention
 SHUNTS Surgical shunts (like a Winter or Glenn shunt) **OTHTX** Other, specify: OTHTXSP **NOTREAT** No specific treatment

12. Are there any other issues that you would like to make us aware of relevant to this patient and his problem with priapism, if a problem exists?

ISSUES

NOSHOW Check this box if the patient did not present for the survey after signing informed consent.

Comprehensive Sickle Cell Centers	NIH Ethnicity/Race	CSCC ID: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> Site: <input type="text"/> <input type="text"/> <input type="text"/>
Epidemiology NIHQ Priapism	Interview Date: <input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/> / <input type="text"/> <input type="text"/> <small>INTDT Day INTMO Month INTYR Year</small>	Identifier acrostic: _____ <small>(for internal use only)</small> Form completed by: <input type="text"/> <input type="text"/> <input type="text"/> <small>INTINIT</small>

Read the following statement and ask the 2 questions of the patient/parent/guardian AT THE END OF THE SURVEY. Read all of the choices before accepting an answer.

Mark the open circle to the left of the question number if the patient declines to answer that question.

The federal government requires that the following questions are asked of all people taking part in a study. Please answer in a way that you feel best describes you.

Q1NA **ETHNIC**
 1. What is your ethnicity? Hispanic or Latino Not Hispanic or Latino

Q2NA
 2. Which of the following racial groups do you consider yourself a part of? **(check all that apply)**

<input type="checkbox"/> AMERIND American Indian/Alaska Native	<input type="checkbox"/> ASIAN Asian	<input type="checkbox"/> AFRAMER Black or African-American
<input type="checkbox"/> HAWAII Native Hawaiian or other Pacific Islander	<input type="checkbox"/> WHITE White	<input type="checkbox"/> Other, specify: RACEOTS _____

Comprehensive Sickle Cell Centers	Survey Instrument Age 5-14	CSCC ID: <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/>
Epidemiology MINR Priapism	Interview Date: INTDA <input style="width:20px; height:20px;" type="text"/> / INTMO <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> / INTYR <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> INTDT Day Month Year	Identifier acoustic: _____ (for internal use only) INTINIT Interviewer's initials: <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/>

- Interview male patients between 5 and 14 years of age, inclusive. Parent or guardian may be present if patient desires. Ensure that there is privacy during conversation. Ask the questions as they are written and record responses on this form. Where instructed, read the possible responses to the patient.
- Text in italics is information for the interviewer and should not be read to the patient/parent/guardian.
- Reminder: As the DSMB has instructed, the patient or parent should be given the honorarium regardless of whether they answer any questions.
- Mark the open circle to the left of the question number if the patient declines to answer that question.

Date informed consent signed: INFCDA / INFCMO / INFCYR
INFCDT Day Month Year

NOANSWER Check this box if the patient chose not to attempt the survey after signing informed consent.

Read the following statement to the patient/parent/guardian before beginning the interview: You are free to refuse to answer any questions you do not wish to answer, and you can stop the interview at any time.

Q1NA
 1. Had you ever heard the word "priapism" before today? PRIAP Yes No Not sure

Read the following statement: Priapism is a painful unwanted erection of the penis that lasts 30 minutes or more.

Q2NA
 2. Before today, had you ever heard of this condition happening in sickle cell disease? Yes No Not sure

Q3NA
 3. Have you [Has your son] ever had a painful unwanted erection of the penis that B4TODAY PAIN Yes No Not sure
 lasted 30 minutes or more?

If answer to #3 is 'No', ask Question 4 and then skip to Question 20.
If answer to #3 is 'Yes' or 'Not sure', the entire questionnaire should be attempted.

Q4NA
 4. Before this study, had a doctor or nurse ever talked with you [you or your son] INFO Yes No Not sure
 about priapism OR provided written information?

Q5ANA
 5a. How many episodes of priapism have you [has your son] had **in the past year**? EPPSTYR
 None 1 2 to 5 6 to 20 More than 20 Don't know

Q5BNA
 5b. How many episodes of priapism did you [your son] have **before the past year**? EPPRIOR
 None 1 2 to 5 6 to 20 More than 20 Don't know

If the patient has only had one episode of priapism (Questions 5a+5b=1), ask Question 6 but skip Questions 7 and 8.
If the patient has had more than one episode of priapism (Questions 5a+5b>1), skip Question 6 and ask Questions 7 and 8.

6a. How old were you [was your son] when the episode of priapism YMOONLY Years ago
Q6ANA happened or how long ago did the episode happen? AGEONLY Years old **or** AGOONLY Months ago

6b. About how many minutes or hours DURMIN Minutes DURHRS Hours DURDAY Days **or** DURUNK Don't know
Q6BNA did the episode of priapism last?

7a. How old were you [was your son] when the **first episode** of YMOFRST Years ago
Q7ANA priapism happened or how long ago did the first episode happen? AGEFRST Years old **or** AGOFRST Months ago

7b. How old were you [was your son] when the **last episode** of YMOLAST Years ago
Q7BNA priapism happened or how long ago did the last episode happen? AGELAST Years old **or** AGOLAST Months ago

8a. About how many minutes or hours does AVGMIN Minutes AVGHRS Hours AVGDAY Days **or** AVGUNK Don't know
Q8ANA an average episode of priapism last? Q8BNA

8b. What about: The shortest episode? SHRTMIN Minutes SHRTHRS Hours SHRTDAY Days **or** SHRTUNK Don't know
Q8CNA

8c. The longest episode? LONGMIN Minutes LONGHRS Hours LONGDAY Days **or** LONGUNK Don't know

8d. The last episode? LASTMIN Minutes LASTHRS Hours LASTDAY Days **or** LASTUNK Don't know
Q8DNA

Epidemiology Priapism

Identifier acrostic: _____
(for internal use only)

Q9ANA
 9a. What time of the day or night does the priapism most often start? *(Read the responses. Check only one.)* **START**

Midnight to 4 in the morning
 4 in the morning to 8 in the morning
 8 in the morning to 4 in the afternoon
 4 in the afternoon to midnight

Q9BNA
 9b. Does priapism most often start when you are [your son is] awake or asleep? *(Question 9c varies with response to this question.)*

	<input type="checkbox"/> Awake AWKASLP	<input type="checkbox"/> Asleep / Wake up with
Q9CNA <input type="radio"/> 9c.	How many hours after waking up does it usually begin? <i>(read the responses)</i> AWAKE	How many hours after going to sleep does it usually begin? <i>(read the responses)</i> ASLEEP
	<input type="checkbox"/> 0-2 hours after waking up <input type="checkbox"/> 2-12 hours after waking up <input type="checkbox"/> > 12 hours after waking up	<input type="checkbox"/> 0-4 hours after going to sleep <input type="checkbox"/> 4-8 hours after going to sleep <input type="checkbox"/> 8-12 hours after going to sleep <input type="checkbox"/> > 12 hours after going to sleep

Q10NA
 10. I am going to read you a list of things that may cause priapism. Do you think that any of these things have caused your [your son's] priapism? *(Read the responses. Check all that apply.)*

- SLEEP** Sleeping
 CONSTIP Constipation
 BREATHE Trouble breathing
 SEXACT Sexual activity (masturbation, intercourse)
 SEXTHTS Sexual thoughts
- ALCOHOL** Alcohol use
 MEDS Medication or drug use (marijuana, herbals, cocaine, antipsychotics) *Specify drug if known:* **MEDSSP**
- CAUSOT** Other cause, *specify:* **CAUSOTS**
 None
 Don't know

Q11NA
 11. Which of the following, if any, do you do to treat the priapism at home? *(Read the responses. Check all that apply.)*

IBUPRO Take non-narcotic pain medication (ibuprofen, acetaminophen, naprosyn)
 CODEINE Take opioids (codeine, hydrocodone)
 MEDOT Other medication, *specify:* **MEDOTS**

URINATE Pee/pass urine/urinate
 SHOWER Take a shower or bath
 REST Rest and relax
 EXRCISE Mild exercise / Get up and walk around

FLUIDS Increase fluids
 TXOTH Other treatment, *specify:* **TXOTHS**
 NOHOMET None

Q12NA
 12. Have you [Has your son] ever been seen at the emergency room, been admitted to the hospital, or seen a doctor **during** an episode of priapism? **DURING**

Yes
 No
 Do not remember

If yes, ask Questions 13 and 14. If 'No' or 'Do not remember' skip to Question 15.

Q13NA
 13. I am going to read you a list of treatments. Tell me which ones you remember being given when you were in the emergency room, hospital, or doctor's office while the painful erection or priapism was still going on. *(Read all responses in the "Treatment" column and check all that apply. For each treatment checked, ask Question 13a and record the response of the priapism.)*

If any treatments are checked, ask Question 13a.

- ORALMED** Treatment
- Pain medication by mouth **ORALSP** →
- IVFLUID**
- Intravenous or IV fluid **FLUIDSP** →
- IVMED**
- Pain medication by vein or shot (IV or IM) **IVMEDSP** →
- TRAN**
- Red blood cell transfusion **TRANSP** →
- NOTREAT**
- None / Does not remember any treatments to end the priapism

Q13ANA
 13a. When you were given the _____ [treatment], what happened to the priapism? *(read responses)*

<input type="checkbox"/> Erection went away/ Pain decreased	<input type="checkbox"/> Stayed about the same/ Do not remember	<input type="checkbox"/> Got worse
<input type="checkbox"/> Erection went away/ Pain decreased	<input type="checkbox"/> Stayed about the same/ Do not remember	<input type="checkbox"/> Got worse
<input type="checkbox"/> Erection went away/ Pain decreased	<input type="checkbox"/> Stayed about the same/ Do not remember	<input type="checkbox"/> Got worse
<input type="checkbox"/> Erection went away/ Pain decreased	<input type="checkbox"/> Stayed about the same/ Do not remember	<input type="checkbox"/> Got worse

Q14NA

14. Do you remember having any of the following surgical procedures to end an episode of priapism either in the emergency department or operating room? (*Read all responses in the "Procedure" column and check all that apply. For each procedure checked, ask Question 14a and record the response of the priapism to the procedure.*)

If any procedures are checked, ask Question 14a.

Procedure

ASPIR

Aspiration, irrigation, or injection
(needle in side of penis to inject medicine or remove blood - may be done in ER or OR)

ASPIRSP

SHUNT

Surgical procedure or shunt
(only done in OR)

SHUNTSP

NOPROC

None / Does not remember any procedures to end the priapism

Q14ANA

14a. When you had the _____ [procedure], what happened to the priapism? (*read the responses*)

Erection went away/
Pain decreased

Stayed about the same/
Do not remember

Got worse

Erection went away/
Pain decreased

Stayed about the same/
Do not remember

Got worse

Q15NA

15. Have you [Has your son] ever received chronic transfusion therapy for any reason (planned red cell transfusion once a month for three months or more)? Yes No Do not remember

CTRAN

Q15ANA

15a. If yes, what happened to your [your son's] priapism during this period of chronic transfusion? (*read the responses*)

Increased

Decreased

Stayed about the same

Was not having episodes of priapism when chronic transfusion was started

Don't know

CTRANYS

Q16NA

16. Have you [Has your son] ever received Hydroxyurea therapy? Yes No Don't know

HYDRX

Q16ANA

16a. If yes, what happened to your [your son's] priapism during the Hydroxyurea therapy? (*read the responses*)

Increased

Decreased

Stayed about the same

Was not having episodes of priapism when Hydroxyurea was started

Don't know

HYDRXYS

17. The next questions are about regular erections, not priapism or painful erections.

a. Do you [Does your son] get erections? Yes No Don't know

b. Do you [Does your son] get erections during the night or wake up with erections? Yes No Don't know

OVRNITE

Q18NA

18. How does the priapism compare to other problems from your [your son's] sickle cell disease? (*read the responses*)

The worst problem

About the same as everything else

Not as bad as other problems

If it is the worst problem, skip to Question 19.

Q18ANA

WORST

18a. Was there ever a time that priapism was the worst problem you [your son] had with sickle cell disease? Yes No Do not remember

Comprehensive Sickle Cell Centers	Survey Instrument Age 5-14	CSCC ID: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>
Epidemiology MINR Priapism		Identifier acrostic: _____ (for internal use only)

Q19NA

19. Is there anything else you want us to know about priapism? _____ **PRIAPSM**

Read the following statement: Thank you for helping us with this study about priapism. Here is a pamphlet with more information. Please let me or any of the clinic staff know if you have any questions about priapism or sickle cell disease. In the future our center may be part of a nationwide study of medicine by mouth to prevent priapism.

Q20NA

20. May we contact you about future studies of priapism? Yes No **CONTACT**

Be sure to complete the NIH Ethnicity/Race CRF for all participants, even those who choose not to attempt the survey.

The following section is for the interviewer and should not be read to the patient/parent/guardian.

21. Who was present during the interview? Patient alone Parent/guardian alone Patient and parent/ guardian
PRESENT

22. Who answered the above questions? Patient alone Parent/guardian alone Patient and parent/ guardian together
ANSWER

23. In what context did the interview occur? **CNTXT**

Urgent Care Visit Routine Visit Health Fair Callback

Chronic Transfusion Hydroxyurea Other, specify: _____ **CNTXTSP**

Please answer demographic questions about yourself. (check those that apply)

GENDER

24. Gender: Male Female

AGE

25. Age: ≤ 30 years 31-49 years ≥ 50 years

INTETH

26. Ethnicity: Hispanic or Latino Not Hispanic or Latino

INTAMIN

27. Race: American Indian / Alaskan Native Native Hawaiian / Other Pacific Islander
(check all that apply)

Asian **INTASIA** White **INTWHITE**

Black or African American **INTOTR** Other, specify: _____ **INTOTRS**

PI Signature: _____ **SIGNTRE**

Signature Date: **SIGDA** / **SIGMO** / **SIGYR**
SIGDT Day Month Year

Comprehensive Sickle Cell Centers	Survey Instrument Age 15-up	CSCC ID: <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> Site: <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/>
Epidemiology ADLT Priapism	Interview Date: INTDA <input style="width:20px; height:20px;" type="text"/> / INTMO <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> / INTYR <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> INTDT Day Month Year	Identifier acoustic: _____ (for internal use only) INTINIT Interviewer's initials: <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/> <input style="width:20px; height:20px;" type="text"/>

- Interview male patients 15 years of age and over. Parent or guardian may be present if patient desires. Ensure that there is privacy during conversation. Ask the questions as they are written and record responses on this form. Where instructed, read the possible responses to the patient.
- Text in italics is information for the interviewer and should not be read to the patient/parent/guardian.
- Reminder: As the DSMB has instructed, the patient or parent should be given the honorarium regardless of whether they answer any questions.
- Mark the open circle to the left of the question number if the patient declines to answer that question.

Date informed consent signed: INFCDA / INFCMO / INFCYR
INFCDT Day Month Year NOANSWER Check this box if the patient chose not to attempt the survey after signing informed consent.

Read the following statement to the patient/parent/guardian before beginning the interview: You are free to refuse to answer any questions you do not wish to answer, and you can stop the interview at any time.

Q1NA 1. Had you ever heard the word "priapism" before today? PRIAP Yes No Not sure

Read the following statement: Priapism is a painful unwanted erection of the penis that lasts 30 minutes or more.

Q2NA 2. Before today, had you ever heard of this condition happening in sickle cell disease? Yes No Not sure

Q3NA 3. Have you ever had a painful unwanted erection of the penis that lasted 30 minutes or more? B4TODAY PAIN Yes No Not sure

If answer to #3 is 'No', ask Question 4 and then skip to Question 20.
If answer to #3 is 'Yes' or 'Not sure', the entire questionnaire should be attempted.

Q4NA 4. Before this study, had a doctor or nurse ever talked with you about priapism OR provided written information? INFO Yes No Not sure

Q5ANA 5a. How many episodes of priapism have you had in the past year? EPPSTYR
 None 1 2 to 5 6 to 20 More than 20 Don't know

Q5BNA 5b. How many episodes of priapism did you have before the past year? EPPRIOR
 None 1 2 to 5 6 to 20 More than 20 Don't know

If the patient has only had one episode of priapism (Questions 5a+5b=1), ask Question 6 but skip Questions 7 and 8.
If the patient has had more than one episode of priapism (Questions 5a+5b>1), skip Question 6 and ask Questions 7 and 8.

Q6ANA 6a. How old were you when the episode of priapism happened or how long ago did the episode happen? AGEONLY Years old AGOONLY Months ago YMOONLY Years ago

Q6BNA 6b. About how many minutes or hours did the episode of priapism last? DURMIN Minutes DURHRS Hours DURDAY Days **or** Don't know DURUNK

Q7ANA 7a. How old were you when the first episode of priapism happened or how long ago did the first episode happen? AGEFRST Years old AGOFRST Months ago YMOFRST Years ago

Q7BNA 7b. How old were you when the last episode of priapism happened or how long ago did the last episode happen? AGELAST Years old AGOLAST Months ago YMOLAST Years ago

Q8ANA 8a. About how many minutes or hours does an average episode of priapism last? AVGMIN Minutes AVGHRS Hours AVGDAY Days **or** Don't know AVGUNK

Q8BNA 8b. What about: The shortest episode? SHRTMIN Minutes SHRTHRS Hours SHRTDAY Days **or** Don't know SHRTUNK

Q8CNA 8c. The longest episode? LONGMIN Minutes LONGHRS Hours LONGDAY Days **or** Don't know LONGUNK

Q8DNA 8d. The last episode? LASTMIN Minutes LASTHRS Hours LASTDAY Days **or** Don't know LASTUNK

Q9ANA
 9a. What time of the day or night does the priapism most often start? *(Read the responses. Check only one.)* **START**

Midnight to 4 in the morning
 4 in the morning to 8 in the morning
 8 in the morning to 4 in the afternoon
 4 in the afternoon to midnight

Q9BNA
 9b. Does priapism most often start when you are awake or asleep? *(Question 9c varies with response to this question.)*

	<input type="checkbox"/> Awake AWKASLP	<input type="checkbox"/> Asleep / Wake up with
Q9CNA <input type="radio"/> 9c. How many hours after waking up does it usually begin? <i>(read the responses)</i> AWAKE	<input type="checkbox"/> 0-2 hours after waking up <input type="checkbox"/> 2-12 hours after waking up <input type="checkbox"/> > 12 hours after waking up	<input type="checkbox"/> 0-4 hours after going to sleep <input type="checkbox"/> 4-8 hours after going to sleep <input type="checkbox"/> 8-12 hours after going to sleep <input type="checkbox"/> > 12 hours after going to sleep

Q10NA
 10. I am going to read you a list of things that may cause priapism. Do you think that any of these things have caused your priapism? *(Read the responses. Check all that apply.)*

- SLEEP** Sleeping
 CONSTIP Constipation
 BREATHE Trouble breathing
 SEXACT Sexual activity (masturbation, intercourse)
 SEXTHTS Sexual thoughts
- ALCOHOL** Alcohol use
 MEDS Medication or drug use (marijuana, herbals, cocaine, antipsychotics) *Specify drug if known:* **MEDSSP**
- CAUSOT** Other cause, *specify:* **CAUSOTS**
 None
 Don't know

Q11NA
 11. Which of the following, if any, do you do to treat the priapism at home? *(Read the responses. Check all that apply.)*

- IBUPRO** Take non-narcotic pain medication (ibuprofen, acetaminophen, naprosyn)
 CODEINE Take opioids (codeine, hydrocodone)
 MEDOT Other medication, *specify:* **MEDOTS**
- URINATE** Pee/pass urine/urinate
 SHOWER Take a shower or bath
 REST Rest and relax
 EXRCISE Mild exercise / Get up and walk around
- FLUIDS** Increase fluids
 TXOTH Other treatment, *specify:* **TXOTHS**
 NOHOMET None

Q12NA
 12. Have you ever been seen at the emergency room, been admitted to the hospital, or seen a doctor **during** an episode of priapism? **DURING**
 Yes
 No
 Do not remember

If yes, ask Questions 13 and 14. If 'No' or 'Do not remember' skip to Question 15.

Q13NA
 13. I am going to read you a list of treatments. Tell me which ones you remember being given when you were in the emergency room, hospital, or doctor's office while the painful erection or priapism was still going on. *(Read all responses in the "Treatment" column and check all that apply. For each treatment checked, ask Question 13a and record the response of the priapism.)*

If any treatments are checked, ask Question 13a.

Treatment

- ORALMED** Pain medication by mouth **ORALSP**
- IVFLUID** Intravenous or IV fluid **FLUIDSP**
- IVMED** Pain medication by vein or shot (IV or IM) **IVMEDSP**
- TRAN** Red blood cell transfusion **TRANSP**
- NOTREAT** None / Does not remember any treatments to end the priapism

Q13ANA
 13a. When you were given the _____ [treatment], what happened to the priapism? *(read responses)*

- | | | |
|--|--|------------------------------------|
| <input type="checkbox"/> Erection went away/
Pain decreased | <input type="checkbox"/> Stayed about the same/
Do not remember | <input type="checkbox"/> Got worse |
| <input type="checkbox"/> Erection went away/
Pain decreased | <input type="checkbox"/> Stayed about the same/
Do not remember | <input type="checkbox"/> Got worse |
| <input type="checkbox"/> Erection went away/
Pain decreased | <input type="checkbox"/> Stayed about the same/
Do not remember | <input type="checkbox"/> Got worse |
| <input type="checkbox"/> Erection went away/
Pain decreased | <input type="checkbox"/> Stayed about the same/
Do not remember | <input type="checkbox"/> Got worse |

Q14NA

14. Do you remember having any of the following surgical procedures to end an episode of priapism either in the emergency department or operating room? (*Read all responses in the "Procedure" column and check all that apply. For each procedure checked, ask Question 14a and record the response of the priapism to the procedure.*)

If any procedures are checked, ask Question 14a.

Procedure

ASPIR

Aspiration, irrigation, or injection
(needle in side of penis to inject medicine or remove blood - may be done in ER or OR)

ASPIRSP

SHUNT

Surgical procedure or shunt
(only done in OR)

SHUNTSP

NOPROC

None / Does not remember any procedures to end the priapism

Q14ANA

14a. When you had the _____ [procedure], what happened to the priapism? (*read the responses*)

Erection went away/
Pain decreased

Stayed about the same/
Do not remember

Got worse

Erection went away/
Pain decreased

Stayed about the same/
Do not remember

Got worse

Q15NA

15. Have you ever received chronic transfusion therapy for any reason (planned red cell transfusion once a month for three months or more)? Yes No Do not remember

CTRAN

Q15ANA

15a. *If yes*, what happened to your priapism during this period of chronic transfusion? (*read the responses*) CTRANYS

Increased

Decreased

Stayed about the same

Was not having episodes of priapism when chronic transfusion was started

Don't know

Q16NA

16. Have you ever received Hydroxyurea therapy? Yes No Don't know

HYDRX

Q16ANA

16a. *If yes*, what happened to your priapism during the Hydroxyurea therapy? (*read the responses*) HYDRXYS

Increased

Decreased

Stayed about the same

Was not having episodes of priapism when Hydroxyurea was started

Don't know

17. The next questions are about regular erections, not priapism or painful erections.

Q17ANA) a. Do you get erections when you want them? WANTED Yes No Don't know

Q17BNA) b. Do you get erections during the night or wake up with erections? Yes No Don't know

Q17CNA) c. Are you able to have sexual intercourse? OVRNITE INTRCRS Yes No Not sexually active

Q17DNA) d. Have you ever been to a doctor for problems with impotence or inability to have an erection when you want one? IMPTNCE Yes No Don't know

Q17ENA) e. Have you ever taken a medicine to help you have an erection? MEDICIN Yes No Don't know

If yes, what is the name of the medication? MEDNAME _____

Q17FNA) f. Do you have a penile implant? IMPLANT Yes No Don't know

Comprehensive Sickle Cell Centers	Survey Instrument Age 15-up	CSCC ID: <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>
Epidemiology ADLT Priapism		Identifier acrostic: _____ <i>(for internal use only)</i>

Q18NA
 18. How does the priapism compare to other problems from your sickle cell disease? (*read the responses*) **PROBLEM**

The worst problem
 About the same as everything else
 Not as bad as other problems

If it is the worst problem, skip to Question 19.

Q18ANA
 18a. Was there ever a time that priapism was the worst problem you had with sickle cell disease? **WORST**

Yes
 No
 Do not remember

Q19NA
 19. Is there anything else you want us to know about priapism? **PRIAPSM**

Read the following statement: Thank you for helping us with this study about priapism. Here is a pamphlet with more information. Please let me or any of the clinic staff know if you have any questions about priapism or sickle cell disease. In the future our center may be part of a nationwide study of medicine by mouth to prevent priapism.

Q20NA
 20. May we contact you about future studies of priapism? Yes No **CONTACT**

Be sure to complete the NIH Ethnicity/Race CRF for all participants, even those who choose not to attempt the survey.

The following section is for the interviewer and should not be read to the patient/parent/guardian.

21. Who was present during the interview? **PRESENT**

Patient alone
 Parent/guardian alone
 Patient and parent/ guardian

22. Who answered the above questions? **ANSWER**

Patient alone
 Parent/guardian alone
 Patient and parent/ guardian together

23. In what context did the interview occur? **CNTXT**

Urgent Care Visit
 Routine Visit
 Health Fair
 Callback

Chronic Transfusion
 Hydroxyurea
 Other, specify: **CNTXTSP** _____

Please answer demographic questions about yourself. (check those that apply)

GENDER
 24. Gender: Male Female

AGE
 25. Age: ≤ 30 years 31-49 years ≥ 50 years

INTETH
 26. Ethnicity: Hispanic or Latino Not Hispanic or Latino

INTAMIN
 27. Race: American Indian / Alaskan Native Native Hawaiian / Other Pacific Islander

Asian **INTASIA**
 Black or African American **INTAFAM** **INTOTR** Other, specify: **INTOTRS** _____

PI Signature: _____ **SIGNTRE**

Signature Date: **SIGDA** / **SIGMO** / **SIGYR**
SIGDT Day Month Year