Walk-PHaSST Annotated CRF - Screening

Phase 1000: Screening

As Needed

Phase 1000:

Phase 1500: Rescreening

Logo	Demographics	{visit.label}
Date of Assessment: DEMO:ASMTDA / DEMO:ASMTMO / DEMO:ASMTYR (Day / Month / Year)		ID: {ID}

1. Date of Birth: DEMO:DOBDA / DEMO:DOBMO / DEMO:DOBYR (Day / Month / Year)

2. Gender: ☐ (DEMO:GENDER) Male ☐ (DEMO:GENDER) Female

Mark the box or boxes that most closely identify the subject's ethnicity and race, as reported by the subject.

3. Ethnicity: ☐ (DEMO:ETHNIC) Hispanic or Latino

☐ (DEMO:ETHNIC) Not Hispanic or Latino

4. Race: (Check all that apply)

☐ (DEMO:RACE1) White

☐ (DEMO:RACE2) Black

Characterize further if possible; otherwise, check "Not otherwise specified"

- ☐ (DEMO:RACE2A) African American (both parents born in America)
- ☐ (DEMO:RACE2B) African Briton (both parents born in United Kingdom)
- ☐ (DEMO:RACE2C) African (both parents born in Africa)
- ☐ (DEMO:RACE2D) Caribbean (both parents born in West Indies)
- ☐ (DEMO:RACE2E) South or Central American (both parents born in South or Central America)
- ☐ (DEMO:RACE2F) Not otherwise specified

☐ (DEMO:RACE3) Asian

☐ (DEMO:RACE4) Native Hawaiian or Pacific Islander

☐ (DEMO:RACE5) American Indian or Alaska Native

☐ (DEMO:RACE6) No Response

☐ (DEMO:RACE7) Unknown

☐ (DEMO:RACE8) Other, specify DEMO:OTH_SP

5. Countries of ancestry: DEMO:COUNTRY ☐ (DEMO:CYUNK) Unknown

6. Has the subject received medical care **outside** the U.S. or United Kingdom for period(s) exceeding 1 year? ☐ (DEMO:OUTCARE) No ☐ (DEMO:OUTCARE) Yes

If **Yes,** for how many years has the subject received medical care **in** the U.S. or United Kingdom? DEMO:CAREYRS

7. Where was the subject born? DEMO:SWHERE

	Mother	Father
8. Where was parent born?	DEMO:MWHERE	DEMO:FWHERE
9. Parent's ethnicity:	☐ (DEMO:METHNIC) Hispanic or Latino ☐ (DEMO:METHNIC) Not Hispanic or Latino ☐ (DEMO:METHNIC) Unknown	☐ (DEMO:FETHNIC) Hispanic or Latino ☐ (DEMO:FETHNIC) Not Hispanic or Latino ☐ (DEMO:FETHNIC) Unknown
10. Parent's race:	(Check all that apply) ☐ (DEMO:MRACE1) White ☐ (DEMO:MRACE2) Black Characterize further if possible; otherwise, check "Not otherwise specified" ☐ (DEMO:MRACE2A) African American (both parents born in America) ☐ (DEMO:MRACE2B) African Briton (both parents born in United Kingdom) ☐ (DEMO:MRACE2C) African (both parents born in Africa) ☐ (DEMO:MRACE2D) Caribbean (both parents born in West Indies) ☐ (DEMO:MRACE2E) South or Central American (both parents born in South or Central America) ☐ (DEMO:MRACE2F) Not otherwise specified ☐ (DEMO:MRACE3) Asian ☐ (DEMO:MRACE4) Native Hawaiian or Pacific Islander ☐ (DEMO:MRACE5) American Indian or Alaska Native	(Check all that apply) ☐ (DEMO:FRACE1) White ☐ (DEMO:FRACE2) Black Characterize further if possible; otherwise, check "Not otherwise specified" ☐ (DEMO:FRACE2A) African American (both parents born in America) ☐ (DEMO:FRACE2B) African Briton (both parents born in United Kingdom) ☐ (DEMO:FRACE2C) African (both parents born in Africa) ☐ (DEMO:FRACE2D) Caribbean (both parents born in West Indies) ☐ (DEMO:FRACE2E) South or Central American (both parents born in South or Central America) ☐ (DEMO:FRACE2F) Not otherwise specified ☐ (DEMO:FRACE3) Asian ☐ (DEMO:FRACE4) Native Hawaiian or Pacific Islander ☐ (DEMO:FRACE5) American Indian or Alaska Native

	(DEMO:MRACE6) No Response (DEMO:MRACE7) Unknown (DEMO:MRACE8) Other, specify DEMO:MOTH_SP	(DEMO:FRACE6) No Response (DEMO:FRACE7) Unknown (DEMO:FRACE8) Other, specify DEMO:FOTH_SP
11. Is parent alive?	(DEMO:MALIVE) No (DEMO:MALIVE) Yes	(DEMO:FALIVE) No (DEMO:FALIVE) Yes
If **No,** cause of death:	DEMO:MCAUSE	DEMO:FCAUSE
12. Diseases and disorders on parent's side of family:	DEMO:MDIS	DEMO:FDIS

13. Number of full siblings: DEMO:FULLSIB (DEMO:FULLUNK) Unknown

14. Number of half siblings: DEMO:HALFSIB (DEMO:HALFUNK) Unknown

Add a Sibling Record for each full and half sibling.

Relationship to subject	Year of Birth	Have Sickle Cell Trait?	Have Sickle Cell Disease?	Living?	Date of Death	Cause of Death	
(SIBL:RELATE) Full sibling (SIBL:RELATE) Half sibling	SIBL:YOB	(SIBL:SCT) No (SIBL:SCT) Yes (SIBL:SCT) Unknown	(SIBL:SCD) No (SIBL:SCD) Yes (SIBL:SCD) Unknown	(SIBL:LIVING) No (SIBL:LIVING) Yes	SIBL:DODDA / SIBL:DODMO / SIBL:DODYR Day Month Year	SIBL:CAUSE	Remo

Add Sibling Record

Comments for page:

DEMO:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo	**Medical History** **Part 1: Diagnosis, Transfusion, Reproductive & Social Histories**	**{visit.label}**
Date of Assessment: MDX1:ASMTDA / MDX1:ASMTMO / MDX1:ASMTYR Day Month Year		**ID: {ID}**

A. Study Diagnosis History

1. Sickle Cell Genotype:
- ☐ (MDX1:SCGENO) SB^{0} (thalassemia)
- ☐ (MDX1:SCGENO) SB^{+} (thalassemia)
- ☐ (MDX1:SCGENO) $SB^{(thalassemia)}$, not otherwise specified
- ☐ (MDX1:SCGENO) SC
- ☐ (MDX1:SCGENO) SD
- ☐ (MDX1:SCGENO) SS
- ☐ (MDX1:SCGENO) Other, specify:

MDX1:SCGENSP

2. Pulmonary hypertension, date of diagnosis: MDX1:PHDA / MDX1:PHMO / MDX1:PHYR (Day Month Year) ☐ (MDX1:PHNA) Not applicable

B. Transfusion History

1. Total number of transfusions in lifetime: ☐ (MDX1:TRANTOT) 0 ☐ (MDX1:TRANTOT) 1-5 ☐ (MDX1:TRANTOT) 6-20 ☐ (MDX1:TRANTOT) 21-100 ☐ (MDX1:TRANTOT) >100

2. Is subject on chronic transfusion therapy? ☐ (MDX1:THRPY) No ☐ (MDX1:THRPY) Yes

If **Yes,** date started: MDX1:THRPYDA / MDX1:THRPYMO / MDX1:THRPYYR (Day Month Year)

If subject has had transfusion:

3. Date of last transfusion: MDX1:TRANDA / MDX1:TRANMO / MDX1:TRANYR (Day Month Year)

4. Type of transfusion: ☐ (MDX1:TRANTYP) Simple ☐ (MDX1:TRANTYP) Exchange ☐ (MDX1:TRANTYP) Other

5. Number of units transfused: MDX1:UNITS

6. Previous transfusion reactions:

- ☐ (MDX1:REAC1) None
- ☐ (MDX1:REAC2) Allergic (fever, urticaria, chills, etc.)
- ☐ (MDX1:REAC3) Alloimmunization (antibodies to transfused red cells)
- ☐ (MDX1:REAC4) Febrile (fever, chills)
- ☐ (MDX1:REAC5) Hemolytic
- ☐ (MDX1:REACOTH) Other, specify: MDX1:REACSP

C. Reproductive History, Female

☐ (MDX1:NOTAPP) Not Applicable

1. Status:

☐ (MDX1:STATUS) Pre-menarche

☐ (MDX1:STATUS) Post-menarche/pre-menopausal, specify:

Age of menarche: MDX1:AGEMEN1

Cycle length: MDX1:CYCLE days

Is cycle regular? ☐ (MDX1:REGULAR) No ☐ (MDX1:REGULAR) Yes

☐ (MDX1:STATUS) Post-menopausal, specify:

Age of menarche: MDX1:AGEMEN2

Age at onset of menopause: MDX1:ONSET

Last menstrual period: MDX1:LMPMO / MDX1:LMPYR

Month Year

2. Number of pregnancies (if female): MDX1:PREG

3. Number of live births (if female): MDX1:BIRTHS

D. Social History

1. Smoking History:

- ☐ (MDX1:SMK) None
- ☐ (MDX1:SMK) Current smoker
- ☐ (MDX1:SMK) Former smoker

If **Current** or **Former smoker:**

Year started: MDX1:SMKYST

Maximum packs/day: MDX1:SMKPKS

Year stopped **(if Former smoker):** MDX1:SMKYSP

2. Alcohol History:

- ☐ (MDX1:ALC) None
- ☐ (MDX1:ALC) Currently drank alcohol
- ☐ (MDX1:ALC) Formerly drank alcohol

If **Formerly drank** or **Currently drink alcohol:**

Maximum drinks/week: MDX1:ALCNUM

3. Drug Use History:

- ☐ (MDX1:DRUG) None
- ☐ (MDX1:DRUG) Current drug use
- ☐ (MDX1:DRUG) Former drug use

If **Former** or **Current drug use,** specify:

- ☐ (MDX1:AMP) Amphetamines
- ☐ (MDX1:COC) Cocaine
- ☐ (MDX1:HER) Heroin
- ☐ (MDX1:MAR) Marijuana
- ☐ (MDX1:DRGOTH) Other, specify:

MDX1:DRGSP

E. Data Collection

1. Indicate how the information reported on this form was collected:

- ☐ (MDX1:COLL) All or most per subject (or parent) report only; not confirmed via medical record
- ☐ (MDX1:COLL) All or most confirmed via medical record
- ☐ (MDX1:COLL) Other, specify: MDX1:COLLSP

Comments for page:

MDX1:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

Logo

Medical History

Part 2: Surgical & Disease Histories

ID:
{visit.label}

Date of Assessment: MDX2:ASMTDA / MDX2:ASMTMO / MDX2:ASMTYR
Day Month Year

A. Surgical History

Has the subject had any of the following surgical procedures?

(If the subject has had the same surgery more than once, record the year of the most recent and provide details in Comment field.)

Procedure	No	Yes	Unknown	Year Performed	Comment/Complications
1. Tonsillectomy/Adenoidectomy	(MDX2:TON)	(MDX2:TON)	(MDX2:TON)	MDX2:TON_YR	MDX2:TON_C
2. Splenectomy	(MDX2:SPL)	(MDX2:SPL)	(MDX2:SPL)	MDX2:SPL_YR	MDX2:SPL_C
3. Cholecystectomy	(MDX2:CHL)	(MDX2:CHL)	(MDX2:CHL)	MDX2:CHL_YR	MDX2:CHL_C
4. Hip Core Procedure	(MDX2:HCP)	(MDX2:HCP)	(MDX2:HCP)	MDX2:HCP_YR	MDX2:HCP_C
5. Hip Replacement	(MDX2:HR)	(MDX2:HR)	(MDX2:HR)	MDX2:HR_YR	MDX2:HR_C
6. Laser Procedure of the Eye(s)	(MDX2:LPE)	(MDX2:LPE)	(MDX2:LPE)	MDX2:LPE_YR	MDX2:LPE_C
7. Vitrectomy	(MDX2:VIT)	(MDX2:VIT)	(MDX2:VIT)	MDX2:VIT_YR	MDX2:VIT_C
8. Insertion of a Permanent Indwelling Line	(MDX2:IPL)	(MDX2:IPL)	(MDX2:IPL)	MDX2:IPL_YR	MDX2:IPL_C
9. Removal of a Permanent Indwelling Line	(MDX2:RPL)	(MDX2:RPL)	(MDX2:RPL)	MDX2:RPL_YR	MDX2:RPL_C
10. Penile Implant	(MDX2:PEN)	(MDX2:PEN)	(MDX2:PEN)	MDX2:PEN_YR	MDX2:PEN_C
11. Other		(MDX2:SUR_OTH)			
If Other is **Yes,** add a Surgery Record for each additional procedure:					

(If the subject has had the same surgery more than once, record the year of the most recent and provide details in Comment field.)

Procedure	Year Performed	Comment/Complications	Remove
MXSG:PROC	MXSG:PROC_YR	MXSG:PROC_C	

Add Surgery Record

B. Diseases/Disorders/Ailments History

Does the subject report having now or in the past any of the following?
(Check all that apply)

	No	Yes
Muscle, Bone or Joint Problems If **Yes,** check all that apply:	☐ (MDX2:MBJ)	☐ (MDX2:MBJ)
1. Hip complications		☐ (MDX2:M_HIP)
2. Was the hip complication avascular necrosis?		☐ (MDX2:M_AVH)
3. Shoulder complication		☐ (MDX2:M_SHL)
4. Was the shoulder complication avascular necrosis?		☐ (MDX2:M_AVS)
5. Dactylitis (Hand Foot Syndrome)		☐ (MDX2:M_DAC)
6. Leg ulcers		☐ (MDX2:M_ULC)
7. Osteomyelitis (acute or chronic)/Bone marrow infection		☐ (MDX2:M_OMY)
8. Osteopenia ("thin bones")		☐ (MDX2:M_OSP)
9. Other, specify: MDX2:M_OTSP		☐ (MDX2:M_OTH)
10. Subject unsure what problem is/was		☐ (MDX2:M_UNSU)
	No	**Yes**
Heart Problems If **Yes,** check all that apply:	☐ (MDX2:HEART)	☐ (MDX2:HEART)
11. Heart failure		☐ (MDX2:H_FAIL)
12. Heart attack		☐ (MDX2:H_ATK)
13. Arrhythmia or prolonged irregular heart beats		☐ (MDX2:H_ARH)
14. Enlarged (big) heart		☐ (MDX2:H_ENL)
15. Cardiomyopathy or "weak heart"		☐ (MDX2:H_CMY)
16. Heart valve problems		☐ (MDX2:H_HVP)
17. High blood pressure/hypertension		☐ (MDX2:H_HYP)
18. Other, specify: MDX2:H_OTSP		☐ (MDX2:H_OTH)
19. Subject unsure what problem is/was		☐ (MDX2:H_UNSU)
	No	**Yes**
Kidney/Urinary/Genital Problems If **Yes,** check all that apply:	☐ (MDX2:KIDNEY)	☐ (MDX2:KIDNEY)
20. Chronic renal (kideny) failure		☐ (MDX2:K_CRF)
21. Pyelonephritis or infection in the kidney		☐ (MDX2:K_PYL)
22. Acute renal (kidney) failure		☐ (MDX2:K_ARF)
23. Chronic Renal Insufficiency		☐ (MDX2:K_CRI)
24. Erectile Dysfunction or impotence		☐ (MDX2:K_EDI)
25. Hematuria or "blood in urine"		☐ (MDX2:K_HEM)
26. Priapism or painful prolonged penile erection		☐ (MDX2:K_PRI)
27. Proteinuria or Nephrotic Syndrome/"protein or albumin in the urine"		☐ (MDX2:K_PROT)
29. Other, specify: MDX2:K_OTSP		☐ (MDX2:K_OTH)
30. Subject unsure what problem is/was		☐ (MDX2:K_UNSU)
	No	**Yes**
Liver Problems		

If **Yes,** check all that apply:	☐ (MDX2:LIVER)	☐ (MDX2:LIVER)
31. Gallbladder disease		☐ (MDX2:L_GALL)
32. Cirrhosis of the liver/hepatic cirrhosis		☐ (MDX2:L_CIRR)
33. Liver failure/hepatic failure		☐ (MDX2:L_FAIL)
34. Liver fibrosis/hepatic fibrosis		☐ (MDX2:L_FIB)
35. Hepatitis, type A		☐ (MDX2:L_HEPA)
36. Hepatitis, type B		☐ (MDX2:L_HEPB)
37. Hepatitis, type C		☐ (MDX2:L_HEPC)
38. Hepatitis, unspecified		☐ (MDX2:L_HEPU)
39. Hepatic sequestration (suddenly enlarged and painful liver, blamed on sickle cell)		☐ (MDX2:L_SEQ)
40. Intrahepatic cholestasis/"bile sludge in the liver"		☐ (MDX2:L_IC)
41. Cholecystitis or gallbladder infection		☐ (MDX2:L_CHCY)
42. Gallstones/cholelithiasis/sludge		☐ (MDX2:L_CHLS)
43. Pancreatitis or inflammation of the pancreas		☐ (MDX2:L_PAN)
44. Transfusional hemosiderosis/"iron in the liver"		☐ (MDX2:L_TH)
45. Other, specify: MDX2:L_OTSP		☐ (MDX2:L_OTH)
46. Subject unsure what problem is/was		☐ (MDX2:L_UNSU)
	No	**Yes**
Spleen Problems If **Yes,** check all that apply:	☐ (MDX2:SPLEEN)	☐ (MDX2:SPLEEN)
47. Splenic infarction		☐ (MDX2:S_INF)
48. Splenomegaly/enlarged spleen		☐ (MDX2:S_SPMG)
49. Chronic hypersplenism/ "spleen has been big for a long time; blood counts may be low because of it"		☐ (MDX2:S_HYPER)
50. Splenic sequestration (sudden enlarged spleen)		☐ (MDX2:S_SEQ)
51. Other, specify: MDX2:S_OTSP		☐ (MDX2:S_OTH)
52. Subject unsure what problem is/was		☐ (MDX2:S_UNSU)
	No	**Yes**
Lung Disease/Problems If **Yes,** check all that apply:	☐ (MDX2:LUNG)	☐ (MDX2:LUNG)
53. Obstructive sleep apnea		☐ (MDX2:P_OSA)
54. Chronic lung disease		☐ (MDX2:P_CLD)
55. Asthma/wheezing/reactive airway		☐ (MDX2:P_ASTH)
56. Pneumonia/acute chest syndrome		☐ (MDX2:P_PNEU)
57. Chronic obstructive lung disease (COPD)/emphysema		☐ (MDX2:P_COPD)
58. Chronic restrictive lung disease/pulmonary fibrosis		☐ (MDX2:P_CRPD)
59. Pulmonary embolism (blood clot to the lung)		☐ (MDX2:P_PE)
60. Pulmonary hypertension		☐ (MDX2:P_PH)
61. Other, specify: MDX2:P_OTSP		☐ (MDX2:P_OTH)
62. Subject unsure what problem is/was		☐ (MDX2:P_UNSU)
	No	**Yes**
Neurological Problems If **Yes,** check all that apply:	☐ (MDX2:NEURO)	☐ (MDX2:NEURO)
63. Seizure		☐ (MDX2:N_SZR)
64. Stroke – hemorrhagic "bleeding in brain"		☐ (MDX2:N_STRH)
65. Stroke – infarct "blocked blood flow to brain"		☐ (MDX2:N_STRI)

66. Stroke – a "silent stroke" seen only on CAT scan or MRI		☐ (MDX2:N_SCI)
67. Elevated transcranial doppler (TCD) velocities		☐ (MDX2:N_TCD)
68. Transient ischemic attack (TIA)/"temporary stroke"		☐ (MDX2:N_TIA)
69. Aneurysm, or balloon-like swelling in blood vessels in brain		☐ (MDX2:N_ANE)
70. Peripheral neuropathy (numbness or tingling, not due to previous stroke)		☐ (MDX2:N_NPTHY)
71. Headache – chronic		☐ (MDX2:N_HDC)
72. Headache – migraine		☐ (MDX2:N_HDM)
73. Memory problems		☐ (MDX2:N_MEM)
74. Depression		☐ (MDX2:N_DEP)
75. Other, specify: MDX2:N_OTSP		☐ (MDX2:N_OTH)
76. Subject unsure what problem is/was		☐ (MDX2:N_UNSU)
	No	**Yes**
Blood Problems, Other Than Sickle Cell If **Yes,** check all that apply:	☐ (MDX2:BLOOD)	☐ (MDX2:BLOOD)
77. Aplastic episode/red blood cell count (or all blood cells counts) severely low		☐ (MDX2:B_APL)
78. Immune and non-immune hemolysis/hyperhemolysis		☐ (MDX2:B_HEMO)
79. Other anemia (not related to sickle cell)		☐ (MDX2:B_ANEM)
80. Low platelets, not due to medication		☐ (MDX2:B_LPT)
81. Low white count, not due to medication		☐ (MDX2:B_LWBC)
82. Other, specify: MDX2:B_OTSP		☐ (MDX2:B_OTH)
83. Subject unsure what problem is/was		☐ (MDX2:B_UNSU)
	No	**Yes**
Infections If **Yes,** check all that apply:	☐ (MDX2:INFECT)	☐ (MDX2:INFECT)
84. Sepsis, "overwhelming blood infection" pneumococcal		☐ (MDX2:I_SEPP)
85. Sepsis, "overwhelming blood infection" other than pneumococcal		☐ (MDX2:I_SEPO)
86. Bacteremia, bacteria in bloodstream (often associated with indwelling catheters)		☐ (MDX2:I_BACT)
87. Meningitis		☐ (MDX2:I_MEN)
88. Other, specify: MDX2:I_OTSP		☐ (MDX2:I_OTH)
89. Subject unsure what problem is/was		☐ (MDX2:I_UNSU)
	No	**Yes**
Other Diseases/Ailments If **Yes,** check all that apply:	☐ (MDX2:OTHDIS)	☐ (MDX2:OTHDIS)
90. Diabetes		☐ (MDX2:O_DIAB)
91. Lupus (SLE)		☐ (MDX2:O_SLE)
92. Rheumatoid arthritis		☐ (MDX2:O_RA)
93. Retinopathy		☐ (MDX2:O_RET)
94. Acute multi-organ failure		☐ (MDX2:O_AMOF)
95. Iron overload		☐ (MDX2:O_IRON)
96. Has iron overload ever been assessed by liver biopsy? ☐ (MDX2:BIOP) No ☐ (MDX2:BIOP) Yes ☐ (MDX2:BIOP) Unknown If **Yes,** result of most recent assessment:		

Assessment	Specimen Date	Result
Liver Biopsy	MDX2:BIOPDA (Day) / MDX2:BIOPMO (Month) / MDX2:BIOPYR (Year)	MDX2:BIOPRS mg Fe/g Dry Weight

97. Vitamin D deficiency		☐ (MDX2:O_VITD)
98. Cancer, describe: MDX2:O_CANSP		☐ (MDX2:O_CAN)
99. Other, specify: MDX2:O_OTSP		☐ (MDX2:O_OTH)
100. Subject unsure what problem is/was		☐ (MDX2:O_UNSU)

C. Diagnostic Tests

Has this subject ever had any of the following diagnostic tests performed: MRI (head), MRA (head), Transcranial Doppler (TCD), Echocardiogram, Pulmonary Function Testing, or EKG?

☐ (MDX2:DIAGTST) No ☐ (MDX2:DIAGTST) Yes

If **Yes,** add a Test Record for each test ever performed:

Test	Test Date	Result	Remove
☐ (DGTS:TYPE) MRI, head ☐ (DGTS:TYPE) MRA, head ☐ (DGTS:TYPE) Transcranial Doppler (TCD) ☐ (DGTS:TYPE) Echocardiogram ☐ (DGTS:TYPE) Pulmonary Function Testing ☐ (DGTS:TYPE) EKG	DGTS:TESTDA / DGTS:TESTMO / DGTS:TESTYR Day Month Year	☐ (DGTS:RESULT) Normal ☐ (DGTS:RESULT) New Abnormal ☐ (DGTS:RESULT) Repeated Abnormal ☐ (DGTS:RESULT) Equivocal	
Comment: DGTS:COMMENT			

Add Test Record

D. Data Collection

Indicate how the information reported on this form was collected:

☐ (MDX2:COLL) All or most per subject (or parent) report only; not confirmed via medical record

☐ (MDX2:COLL) All or most confirmed via medical record

☐ (MDX2:COLL) Other, specify: MDX2:COLLSP

Comments for page:

MDX2:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo

Medical History

Part 3: Medications and Pain Histories

ID:
{visit.label}

Date of Assessment: MDX3:ASMTDA / MDX3:ASMTMO / MDX3:ASMTYR
Day Month Year

A. Medications History

Medication	Currently Using	Used in the Past	Cumulative Lifetime Use
1. Anticoagulation medication	(MDX3:ACG)	(MDX3:ACG)	
2. Anticonvulsants, specify: MDX3:ACVSP	(MDX3:ACV)	(MDX3:ACV)	
3. Antidepressants, specify: MDX3:ADPSP	(MDX3:ADP)	(MDX3:ADP)	
4. Erythropoietin/Darbepoietin	(MDX3:ERY)	(MDX3:ERY)	(MDX3:ERYC) <1 year (MDX3:ERYC) 1-5 years (MDX3:ERYC) >5 years
5. Folic Acid	(MDX3:FOL)	(MDX3:FOL)	
6. Hydroxyurea (Hydroxycarbamide)	(MDX3:HYD)	(MDX3:HYD)	(MDX3:HYDC) <1 year (MDX3:HYDC) 1-5 years (MDX3:HYDC) >5 years
7. Inhalers	(MDX3:INHAL)	(MDX3:INHAL)	
8. Other anti-sickling agents, specify: MDX3:OASASP	(MDX3:OASA)	(MDX3:OASA)	
9. Iron chelation therapy (e.g., Desferal, Exjade, etc.) (MDX3:IRONTYP) Desferal (Deferoxamine) (MDX3:IRONTYP) Exjade (Deferasirox) (MDX3:IRONTYP) Other, specify: MDX3:IRONSP	(MDX3:IRON)	(MDX3:IRON)	(MDX3:IRONC) <1 year (MDX3:IRONC) 1-5 years (MDX3:IRONC) >5 years
10. Oxygen at home	(MDX3:OXY)	(MDX3:OXY)	(MDX3:OXYC) <1 year (MDX3:OXYC) 1-5 years (MDX3:OXYC) >5 years
11. Prophylactic penicillin or other antibiotics	(MDX3:PEN)	(MDX3:PEN)	
Medication	**Currently Using**	**Used in the Past**	**Cumulative Lifetime Use**
12. Pain Medications: Narcotics, daily for 30+ days			
a). Codeine	(MDX3:COD)	(MDX3:COD)	
b). Demerol (Pethidine)	(MDX3:DEM)	(MDX3:DEM)	

c). Dilaudid (Hydromorphone)	☐ (MDX3:DIL)	☐ (MDX3:DIL)	
d). Morphine	☐ (MDX3:MOR)	☐ (MDX3:MOR)	
e). Oxycodone	☐ (MDX3:OXCD)	☐ (MDX3:OXCD)	
f). Oxycontin (Oxycodone hydrochloride)	☐ (MDX3:OXCT)	☐ (MDX3:OXCT)	
g). Percocet (Oxycodone w/ Paracetamol)	☐ (MDX3:PERC)	☐ (MDX3:PERC)	
h). Tylenol 3 (Paracetamol w/ Codein No. 3)	☐ (MDX3:TYL3)	☐ (MDX3:TYL3)	
i). Vicodin (Hydrocodone w/ Paracetamol)	☐ (MDX3:VIC)	☐ (MDX3:VIC)	
j). Methadone	☐ (MDX3:METH)	☐ (MDX3:METH)	
Medication	**Currently Using**	**Used in the Past**	**Cumulative Lifetime Use**
13. Pain Medications: NSAIDs daily for 30+ days (e.g., Aleve [Naproxen], Ibuprofen, Motrin, etc.), specify: MDX3:NSAIDSP	☐ (MDX3:NSAID)	☐ (MDX3:NSAID)	☐ (MDX3:NSAIDC) <1 year ☐ (MDX3:NSAIDC) 1-5 years ☐ (MDX3:NSAIDC) >5 years
14. Other Pain Medications daily for 30+ days (e.g., Gabapentin, Nortriptyline, Elavil, etc.), specify: MDX3:OPMSP	☐ (MDX3:OPM)	☐ (MDX3:OPM)	
15. Pulmonary Hypertension Therapy			
a). Endothelin-receptor antagonist (e.g., Bosentan)	☐ (MDX3:ERA)	☐ (MDX3:ERA)	
b). PDE-5 inhibitor (e.g., Sildenafil)	☐ (MDX3:PDE)	☐ (MDX3:PDE)	
c). Prostacyclin	☐ (MDX3:PCY)	☐ (MDX3:PCY)	
16. Heart/BloodPressure Medications			
a). ACE inhibitors (e.g., Lisinopril, Ramipril, Enalapril, etc.)	☐ (MDX3:ACE)	☐ (MDX3:ACE)	
b). Beta blockers (e.g., Atenolol, Sotalol, etc.)	☐ (MDX3:BETA)	☐ (MDX3:BETA)	
c). Calcium chanel blockers (e.g., Diltiazem, Cardizem, Varapamil, Amlodipine, etc.)	☐ (MDX3:CCB)	☐ (MDX3:CCB)	
d). Diuretics (e.g., Hydrochlorothiazide, Lasix [Furosemide], etc.)	☐ (MDX3:DIUR)	☐ (MDX3:DIUR)	
e). Vasodilators (e.g., Isordil, Isosorbide, prazosin, minipress, cardura)	☐ (MDX3:VASO)	☐ (MDX3:VASO)	
f). Other, specify MDX3:PHT_SP	☐ (MDX3:PHT_OTH)	☐ (MDX3:PHT_OTH)	
Medication	**Currently Using**	**Used in the Past**	**Cumulative Lifetime Use**
17. Renal replacement therapy (e.g., dialysis or kidney transplant), specify: MDX3:RRT_SP	☐ (MDX3:RRT)	☐ (MDX3:RRT)	
18. Other alternative therapies (herbal treatments, antioxidants, vitamin C, etc.), specify: MDX3:ALT_SP	☐ (MDX3:ALT)	☐ (MDX3:ALT)	

19. Previous medication reactions:

(Check all that apply)

- ☐ (MDX3:REAC1) None
- ☐ (MDX3:REAC2) Allergic (fever, urticaria, chills, etc.)
- ☐ (MDX3:REAC3) Alloimmunization (antibodies to transfused red cells)
- ☐ (MDX3:REAC4) Febrile (fever, chills)
- ☐ (MDX3:REAC5) Hemolytic
- ☐ (MDX3:REACOTH) Other, specify: MDX3:REACSP

List medications that caused reactions: MDX3:MEDS

B. Sickle Cell Pain History

Acute Pain

1. Location: (Check all that apply)
☐ (MDX3:AC_LOC1) Arms ☐ (MDX3:AC_LOC2) Chest ☐ (MDX3:AC_LOC3) Joints ☐ (MDX3:AC_LOC4) Neck
☐ (MDX3:AC_LOC5) Back ☐ (MDX3:AC_LOC6) Head ☐ (MDX3:AC_LOC7) Legs
☐ (MDX3:AC_LOCO) Other, specify:
MDX3:AC_LOCS

2. Typical pain rating on 1-10 scale: MDX3:AC_RATE

3. Quality/type of pain: MDX3:AC_QUAL

4. Treatment: (Check all that apply)
☐ (MDX3:AC_TR1) Medication ☐ (MDX3:AC_TR2) Non-Drug Therapy ☐ (MDX3:AC_TR3) Accupuncture
☐ (MDX3:AC_TR4) Physical Therapy ☐ (MDX3:AC_TR5) Alternative Therapy ☐ (MDX3:AC_TR6) Hypnosis
☐ (MDX3:AC_TRO) Other, specify:
MDX3:AC_TRS

	Mild	Moderate	Severe	Extremely Severe
5. Number of pain crises (events) in last week:	MDX3:PNC_WMI	MDX3:PNC_WMO	MDX3:PNC_WSE	MDX3:PNC_WEX
6. Number of pain crises (events) in last month:	MDX3:PNC_MMI	MDX3:PNC_MMO	MDX3:PNC_MSE	MDX3:PNC_MEX
7. Number of pain crises (events) in last year:	MDX3:PNC_YMI	MDX3:PNC_YMO	MDX3:PNC_YSE	MDX3:PNC_YEX

Mild = May or may not have required pain medicine, but did not prevent normal daily activity
Moderate = Required medications and caused significant changes in daily activities (i.e., missing work)
Severe = Went to ER but was not admitted
Extremely Severe = Admitted to the hospital

Chronic Pain

8. Does subject also have chronic pain (present all or most of the time)? ☐ (MDX3:CHRON) No ☐ (MDX3:CHRON) Yes

If **Yes:**

a. Location: (Check all that apply)
☐ (MDX3:CH_LOC1) Arms ☐ (MDX3:CH_LOC2) Chest ☐ (MDX3:CH_LOC3) Joints ☐ (MDX3:CH_LOC4) Neck
☐ (MDX3:CH_LOC5) Back ☐ (MDX3:CH_LOC6) Head ☐ (MDX3:CH_LOC7) Legs
☐ (MDX3:CH_LOCO) Other, specify:
MDX3:CH_LOCS

b. Typical pain rating on 1-10 scale: MDX3:CH_RATE

c. Quality/type of pain: MDX3:CH_QUAL

d. Treatment: (Check all that apply)
☐ (MDX3:CH_TR1) Medication ☐ (MDX3:CH_TR2) Non-Drug Therapy ☐ (MDX3:CH_TR3) Accupuncture
☐ (MDX3:CH_TR4) Physical Therapy ☐ (MDX3:CH_TR5) Alternative Therapy ☐ (MDX3:CH_TR6) Hypnosis
☐ (MDX3:CH_TRO) Other, specify:
MDX3:CH_TRS

C. Data Collection

Indicate how the information reported

on this form was collected:

(MDX3:COLL) All or most per subject (or parent) report only; not confirmed via medical record

(MDX3:COLL) All or most confirmed via medical record

(MDX3:COLL) Other, specify: MDX3:COLLSP

Comments for page:

MDX3:COMM

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Logo	Physical Examination	{visit.label}
Date of Assessment: PHEX:ASMTDA / PHEX:ASMTMO / PHEX:ASMTYR (Day / Month / Year)		ID: {ID}

1. Temperature: PHEX:TEMP °C

2. Heart rate: PHEX:HRATE beats/min

3 Oxygen Saturation PHEX:O2 %, measured on: ☐ (PHEX:AIRO2) Air ☐ (PHEX:AIRO2) O_2

If **O_2**, flow rate: PHEX:O2FLOW L/m

4. Respiratory rate: PHEX:RESP breaths/min

5. Sitting blood pressure: (systolic/diastolic) PHEX:SYSBP / PHEX:DIABP mmHg

6. Weight: PHEX:WEIGHT kg

7. Height: PHEX:HEIGHT cm

8. Body surface area[1]: PHEX:BSA m^2 (round to 2 decimal places)

Category	Status
9. General Appearance	Appearance: ☐ (PHEX:APP) Well appearing ☐ (PHEX:APP) Ill appearing
	Weight: ☐ (PHEX:APPWT) Normal/well nourished ☐ (PHEX:APPWT) Overweight/obese ☐ (PHEX:APPWT) Malnourished/thin
	Comment/ other findings or abnormalities:

PHEX:APPCM

10. HEENT

Scleral icterus: ☐ (PHEX:HEENTSI) None ☐ (PHEX:HEENTSI) Mild ☐ (PHEX:HEENTSI) Moderate

Tonsillar hypertrophy: ☐ (PHEX:HEENTTH) Present ☐ (PHEX:HEENTTH) Absent

Hypopharynx: ☐ (PHEX:HEENTHP) Narrowed ☐ (PHEX:HEENTHP) Normal

`

Comment/ other findings or abnormalities: PHEX:HEENTCM

11. Neurologic – Check all that apply:

☐ (PHEX:NEUR1) Alert and oriented

☐ (PHEX:NEUR2) Normal strength

☐ (PHEX:NEUR3) Normal tone

☐ (PHEX:NEUR4) Normal gait

☐ (PHEX:NEUR5) Stroke sequelae present, describe:

PHEX:STROKE

Comment/ other findings or abnormalities: PHEX:NEURCM

12. Cardiac Heart Sounds

S1 and S2: ☐ (PHEX:CARS12) Normal

S3: ☐ (PHEX:CARS3) Present

S4: ☐ (PHEX:CARS4) Present

P2: ☐ (PHEX:CARP2) Loud

Other Findings ☐ (PHEX:CAROTH) Yes, describe:

PHEX:CARDES

Rate and rhythm: ☐ (PHEX:CARR) Regular ☐ (PHEX:CARR) Irregular, describe:

PHEX:CARRD

Murmur: ☐ (PHEX:CARM) Normal – S1 and S2 with flow mumur heard best at the left upper sternal border

☐ (PHEX:CARM) Other, describe:

PHEX:CARMD

Jugulovenous distension: ☐ (PHEX:CARJD) Present ☐ (PHEX:CARJD) Absent

Comment/ other findings or abnormalities: PHEX:CARCM

13. Pulmonary

Lungs: ☐ (PHEX:PULCL) Clear to ascultation

☐ (PHEX:PULBC) Bibasilar crackles

☐ (PHEX:PULBW) Wheezes

Comment/ other findings or abnormalities: PHEX:PULCM

14. Gastrointestinal – Check all that apply:

Abdomen: ☐ (PHEX:GASTNR) Normal: Belly soft and non-tender

☐ (PHEX:SPL) Splenomegaly:

Size below costal margin: PHEX:SPLSIZE cm

☐ (PHEX:HEP) Hepatomegaly:

Size below costal margin: PHEX:HEPSIZE cm

☐ (PHEX:GASTOT) Other, specify:

Comment/ other findings or abnormalities: PHEX:GASTCM

15. Extremities & Skin

Nails: ☐ (PHEX:EXTNN) Normal ☐ (PHEX:EXTNC) Clubbing ☐ (PHEX:EXTNH) Hyperpigmentation

Lower Extremities

Edema: ☐ (PHEX:EXTED) None ☐ (PHEX:EXTED) + ☐ (PHEX:EXTED) ++ ☐ (PHEX:EXTED) +++ ☐ (PHEX:EXTED) ++++

Pulses: ☐ (PHEX:EXTPL) Normal ☐ (PHEX:EXTPL) Abnormal

If **Abnormal,** describe:

PHEX:EXTPLD

Ulcers: ☐ (PHEX:EXTLUL) None ☐ (PHEX:EXTLUL) Active ☐ (PHEX:EXTLUL) Healed

If **Active** or **Healed,** describe:

PHEX:EXTLULD

Skin

Hyperpigmentation: ☐ (PHEX:SKINHP) Absent ☐ (PHEX:SKINHP) Present

Rashes or skin lesions: ☐ (PHEX:EXTSL) Present ☐ (PHEX:EXTSL) Absent

If **Present,** describe:

PHEX:EXTSLD

Comment/ other findings or abnormalities

PHEX:EXTCM

Comments for page:

PHEX:COMM

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Logo	Chemistry	{visit.label}
Date of Collection: CHEM:COLLDA (Day) / CHEM:COLLMO (Month) / CHEM:COLLYR (Year)		ID: {ID}

Test	Lab Value	Unit	Clinical Significance
Albumin	CHEM:ALB	☐ (CHEM:ALBUNIT) g/dL ☐ (CHEM:ALBUNIT) g/L	☐ (CHEM:ALBSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:ALBSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:ALBSG) Not clinically significant
Alkaline Phosphatase	CHEM:ALK	U/L (IU/L)	☐ (CHEM:ALKSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:ALKSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:ALKSG) Not clinically significant
ALT	CHEM:ALT	U/L (IU/L)	☐ (CHEM:ALTSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:ALTSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:ALTSG) Not clinically significant
AST	CHEM:AST	U/L (IU/L)	☐ (CHEM:ASTSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:ASTSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:ASTSG) Not clinically significant
CO_2	CHEM:CO2	☐ (CHEM:CO2UNIT) mmol/L ☐ (CHEM:CO2UNIT) kPa	☐ (CHEM:CO2SG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:CO2SG) Clinically significant, but not a new Adverse Event ☐ (CHEM:CO2SG) Not clinically significant
BUN	CHEM:BUN	☐ (CHEM:BUNUNIT) mg/dL ☐ (CHEM:BUNUNIT) g/dL ☐ (CHEM:BUNUNIT) mmol/L	☐ (CHEM:BUNSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:BUNSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:BUNSG) Not clinically significant
Calcium	CHEM:CAL	☐ (CHEM:CALUNIT) mg/dL ☐ (CHEM:CALUNIT) mmol/L	☐ (CHEM:CALSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:CALSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:CALSG) Not clinically significant
Chloride	CHEM:CHL	mmol/L	☐ (CHEM:CHLSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:CHLSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:CHLSG) Not clinically significant
Creatinine	CHEM:CRE	☐ (CHEM:CRUNIT) mg/dL ☐ (CHEM:CRUNIT) µmol/L	☐ (CHEM:CRESG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:CRESG) Clinically significant, but not a new Adverse Event ☐ (CHEM:CRESG) Not clinically significant
LDH	CHEM:LDH	IU/L (U/L)	☐ (CHEM:LDHSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:LDHSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:LDHSG) Not clinically significant
Magnesium	CHEM:MAG	☐ (CHEM:MAGUNIT) mg/dL ☐ (CHEM:MAGUNIT) mmol/L ☐ (CHEM:MAGUNIT) mEq/L	☐ (CHEM:MAGSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:MAGSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:MAGSG) Not clinically significant
Phosphate/Phosphorus	CHEM:PHOS	☐ (CHEM:PUNIT) mg/dL Phosphorus ☐ (CHEM:PUNIT) mmol/L Phosphate	☐ (CHEM:PHOSSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:PHOSSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:PHOSSG) Not clinically significant
Potassium	CHEM:POT	mmol/L	☐ (CHEM:POTSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:POTSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:POTSG) Not clinically significant
Sodium	CHEM:SOD	mmol/L	☐ (CHEM:SODSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:SODSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:SODSG) Not clinically significant
Total Bilirubin	CHEM:TBIL	☐ (CHEM:TBUNIT) mg/dL	☐ (CHEM:TBILSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:TBILSG) Clinically significant, but not a new Adverse Event

		☐ (CHEM:TBUNIT) µmol/L	☐ (CHEM:TBILSG) Not clinically significant
Total Protein	CHEM:TPROT	☐ (CHEM:TPUNIT) g/dL ☐ (CHEM:TPUNIT) g/L	☐ (CHEM:TPROTSG) Clinically significant and a new Adverse Event [1] ☐ (CHEM:TPROTSG) Clinically significant, but not a new Adverse Event ☐ (CHEM:TPROTSG) Not clinically significant

[1]Complete an Adverse Events form

Comments for page:

CHEM:COMM

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Logo	Hematology	{visit.label}
Date of Collection: HEMA:COLLDA (Day) / HEMA:COLLMO (Month) / HEMA:COLLYR (Year)		ID:{ID}

Test	Lab Value	Units	Clinical Significance
Absolute Neutrophil Count (ANC)	HEMA:ANC	x10^{3}cells/µL (x10^{9}cells/L)	☐ (HEMA:ANCSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:ANCSG) Clinically significant but not a new Adverse Event ☐ (HEMA:ANCSG) Not clinically significant
Neutrophils (%)	HEMA:NEUT	%	☐ (HEMA:NEUTSG)Clinically significant and a new Adverse Event[1] ☐ (HEMA:NEUTSG) Clinically significant but not a new Adverse Event ☐ (HEMA:NEUTSG) Not clinically significant
Absolute Reticulocyte Count (ARC)	HEMA:ARC	x10^{3}cells/µL (x10^{9}cells/L)	☐ (HEMA:ARCSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:ARCSG) Clinically significant but not a new Adverse Event ☐ (HEMA:ARCSG) Not clinically significant
Reticulocytes (%)	HEMA:RET	%	☐ (HEMA:RETSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:RETSG) Clinically significant but not a new Adverse Event ☐ (HEMA:RETSG) Not clinically significant
Hematocrit	HEMA:HCT	☐ (HEMA:HCTUNIT) % ☐ (HEMA:HCTUNIT) 1:1	☐ (HEMA:HCTSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:HCTSG) Clinically significant but not a new Adverse Event ☐ (HEMA:HCTSG) Not clinically significant
Hemoglobin	HEMA:HGB	☐ (HEMA:HGBUNIT) g/dL ☐ (HEMA:HGBUNIT) g/L	☐ (HEMA:HGBSG)Clinically significant and a new Adverse Event[1] ☐ (HEMA:HGBSG) Clinically significant but not a new Adverse Event ☐ (HEMA:HGBSG) Not clinically significant
Mean Corpuscular Hemoglobin Concentration (MCHC)	HEMA:MCHC	☐ (HEMA:MCHUNIT) g/dL ☐ (HEMA:MCHUNIT) g/L	☐ (HEMA:MCHCSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:MCHCSG) Clinically significant but not a new Adverse Event ☐ (HEMA:MCHCSG) Not clinically significant
Mean Corpuscular Volume (MCV)	HEMA:MCV	fL	☐ (HEMA:MCVSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:MCVSG)Clinically significant but not a new Adverse Event ☐ (HEMA:MCVSG) Not clinically significant
Platelet Count	HEMA:PLAT	x10^{3}cells/µL (x10^{9}cells/L)	☐ (HEMA:PLATSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:PLATSG) Clinically significant but not a new Adverse Event ☐ (HEMA:PLATSG) Not clinically significant
RBC	HEMA:RBC	x10^{6}cells/µL	☐ (HEMA:RBCSG)Clinically significant and a new Adverse Event[1] ☐ (HEMA:RBCSG) Clinically significant but not a new Adverse Event ☐ (HEMA:RBCSG) Not clinically significant
WBC	HEMA:WBC	x10^{3}cells/µL	☐ (HEMA:WBCSG) Clinically significant and a new Adverse Event[1] ☐ (HEMA:WBCSG) Clinically significant but not a new Adverse Event ☐ (HEMA:WBCSG) Not clinically significant

[1]Complete an Adverse Events form

Comments for page:

HEMA:COMM

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Logo	Urinalysis	{visit.label}
		ID: {ID}

1. Date of collection for urinalysis: URIN:COLLDA / URIN:COLLMO / URIN:COLLYR
Day Month Year

Urine Dipstick Chemical Analysis

2. Is the subject menstruating? ☐ (URIN:MENSES) No ☐ (URIN:MENSES) Yes ☐ (URIN:MENSES) Not Applicable (male subject)

3. pH: URIN:PH

4. Specific Gravity: URIN:SPGRAV

For items 5-7, if dipstick result is positive, record code and/or value

Test	Dipstick Results	Code	Value
5. Glucose	☐ (URIN:GLU) Negative ☐ (URIN:GLU) Positive	☐ (URIN:GLUCODE) Trace ☐ (URIN:GLUCODE) 1+ ☐ (URIN:GLUCODE) 2+ ☐ (URIN:GLUCODE) 3+ ☐ (URIN:GLUCODE) 4+	URIN:GLUVAL mg/dL
6. Protein (Proteinuria)	☐ (URIN:PRO) Negative ☐ (URIN:PRO) Positive	☐ (URIN:PROCODE) Trace ☐ (URIN:PROCODE) 1+ ☐ (URIN:PROCODE) 2+ ☐ (URIN:PROCODE) 3+ ☐ (URIN:PROCODE) 4+	URIN:PROVAL mg/dL
7. Blood	☐ (URIN:BLD) Negative ☐ (URIN:BLD) Positive	☐ (URIN:BLDCODE) Trace ☐ (URIN:BLDCODE) 1+ ☐ (URIN:BLDCODE) 2+ ☐ (URIN:BLDCODE) 3+ ☐ (URIN:BLDCODE) 4+	URIN:BLDVAL Ery/µL

Microscopic Exam

8. Was microscopic exam performed? ☐ (URIN:MICROYN) No ☐ (URIN:MICROYN) Yes

If **Yes,** complete the following:

RBC: ☐ (URIN:RBC) 0-5 ☐ (URIN:RBC) 5-10 ☐ (URIN:RBC) 10-25 ☐ (URIN:RBC) 25-50 ☐ (URIN:RBC) 50+ #/HPF

WBC: ☐ (URIN:WBC) 0-5 ☐ (URIN:WBC) 5-10 ☐ (URIN:WBC) 10-25 ☐ (URIN:WBC) 25-50 ☐ (URIN:WBC) 50+ #/HPF

9. Other abnormal findings on microscopic exam? ☐ (URIN:OMICYN) No ☐ (URIN:OMICYN) Yes

If **Yes,** describe: URIN:OMICDES

Overall Assessment of Urinalysis

10. Overall assessment of urinalysis: ☐ (URIN:OVERALL) Normal ☐ (URIN:OVERALL) Abnormal

If **Abnormal,** is this a new AE? ☐ (URIN:NEWAE) No ☐ (URIN:NEWAE) Yes (If **Yes,** report on Adverse Events form.)

Albumin/Creatinine

11. Date of collection for albumin/creatinine: URIN:ACDA / URIN:ACMO / URIN:ACYR

Day Month Year

12. Albumin/creatinine URIN:ALB : URIN:CREAT **or** URIN:RATIO

albumin (mg/dL) creatinine (mg/dL) ratio

Comments for page:

URIN:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo	HIV Test	{visit.label}
Date of Collection: HIVT:COLLDA / HIVT:COLLMO / HIVT:COLLYR (Day / Month / Year)		ID: {ID}

Result: (HIVT:RESULT) Negative (HIVT:RESULT) Positive

If **Positive,** is subject on protease inhibitor therapy for treatment of HIV? (HIVT:PIT) No (HIVT:PIT) Yes

Comments for page:

HIVT:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo	Pregnancy Test	{visit.label}
Date of Assessment: PREG:ASMTDA / PREG:ASMTMO / PREG:ASMTYR (Day / Month / Year)		ID: {ID}

1. Result:

- ☐ (PREG:RESULT) Negative
- ☐ (PREG:RESULT) Positive
- ☐ (PREG:RESULT) Not Done

If **Not Done,** specify reason:

- ☐ (PREG:REASON) Subject is male
- ☐ (PREG:REASON) Subject is pre-menarche or post-menopausal
- ☐ (PREG:REASON) Subject is surgically or medically sterile
- ☐ (PREG:REASON) Other, specify: PREG:OTH_SP

2. Type of test: ☐ (PREG:TYPE) Serum ☐ (PREG:TYPE) Urine

If **Serum,** HCG: PREG:HCGSYM PREG:HCG mIU/mL (IU/L) Q2:<; >

Comments for page: PREG:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

Logo

Echocardiogram (Local)

{visit.label}

Date of Procedure: ECHO:PROCDA / ECHO:PROCMO / ECHO:PROCYR
Day Month Year

ID: {ID}

1.Time of procedure: ECHO:PROCHH : ECHO:PROCMM
Hr Min (24 hr clock)

2. Blood pressure at time of procedure: ECHO:SYSBP / ECHO:DIABP mmHg
(Systolic / Diastolic)

3. Tricuspid regurgitant jet velocity: ECHO:TRJV m/s **or** (ECHO:TRJVND) Not detectable

4. Estimated right atrial pressure (mmHg): (ECHO:ERAP) 5 (ECHO:ERAP) 10 (ECHO:ERAP) 15 (ECHO:ERAP) 20

5. LV function: (ECHO:LVFUN)Normal (ECHO:LVFUN)Abnormal

6. LV ejection fraction: ECHO:LVEF

	None	Trace	Mild	Mild-Moderate	Moderate	Moderate-Severe	Severe
7. Aortic regurgitation:	(ECHO:AR)	(ECHO:AR)	(ECHO:AR)	(ECHO:AR)	(ECHO:AR)	(ECHO:AR)	(ECHO:AR)
8. Mitral regurgitation:	(ECHO:MR)	(ECHO:MR)	(ECHO:MR)	(ECHO:MR)	(ECHO:MR)	(ECHO:MR)	(ECHO:MR)
9. Tricuspid regurgitation:	(ECHO:TR)	(ECHO:TR)	(ECHO:TR)	(ECHO:TR)	(ECHO:TR)	(ECHO:TR)	(ECHO:TR)

10. Other significant findings:
ECHO:OTHSIG

11. Echo recording bar code(s): ECHO:CODE1 ECHO:CODE2

Comments for page:
ECHO:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo	6-Minute Walk Test	{visit.label}
Date of Assessment: SIXM:ASMTDA / SIXM:ASMTMO / SIXM:ASMTYR (Day / Month / Year)		ID: {ID}

Before Walk

1. Blood pressure (systolic/diastolic): SIXM:SYSPRE / SIXM:DIAPRE mmHg

2. Heart rate: SIXM:HRPRE beats/min

3. O_2 saturation: SIXM:OSPRE %, measured on: ☐ (SIXM:AOPRE) Air ☐ (SIXM:AOPRE) O_2

   If **O_2**, flow rate: SIXM:FRPRE L/m

4. Time walk started (24-hr clock) SIXM:STARTHH : SIXM:STARTMM (Hr Min)

After Walk

5. Blood pressure (systolic/diastolic): SIXM:SYSPOST / SIXM:DIAPOST mmHg

6. Heart rate immediately after: SIXM:HRPOST beats/min

7. O_2 saturation immediately after: SIXM:OSPOST %, measured on: ☐ (SIXM:AOPOST) Air ☐ (SIXM:AOPOST) O_2

   If **O_2**, flow rate: SIXM:FRPOST L/m

8. Distance walked: SIXM:DIST m

9. Did subject stop before 6-minute time limit? ☐ (SIXM:SUBSTOP) No ☐ (SIXM:SUBSTOP) Yes

10. Did subject use oxygen during the test? ☐ (SIXM:USEO2) No ☐ (SIXM:USEO2) Yes

    If **Yes:** Oxygen flow rate: SIXM:FRWALK L/min

    Was oxygen device carried or pushed? ☐ (SIXM:DEVICE) Carried ☐ (SIXM:DEVICE) Pushed

11. Borg dyspnea score:

    ☐ (SIXM:BRGPOST) 0=Nothing at all

    ☐ (SIXM:BRGPOST) 0.5=Very, very slight (just noticeable)

☐ (SIXM:BRGPOST) 1=Very slight

☐ (SIXM:BRGPOST) 2=Slight

☐ (SIXM:BRGPOST) 3=Moderate

☐ (SIXM:BRGPOST) 4=Somewhat severe

☐ (SIXM:BRGPOST) 5=severe

☐ (SIXM:BRGPOST) 6

☐ (SIXM:BRGPOST) 7=Very severe

☐ (SIXM:BRGPOST) 8

☐ (SIXM:BRGPOST) 9=Very, very severe (almost maximum)

☐ (SIXM:BRGPOST) 10=Maximum

---

12. NYHA/WHO classification:

☐ (SIXM:CLASS) Class I	Patients with pulmonary hypertension but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnea or fatigue, chest pain, or near syncope.
☐ (SIXM:CLASS) Class II	Patients with pulmonary hypertension resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnea or fatigue, chest pain, or near syncope.
☐ (SIXM:CLASS) Class III	Patients with pulmonary hypertension resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary physical activity causes undue dyspnea or fatigue, chest pain, or near syncope.
☐ (SIXM:CLASS) Class IV	Patients with pulmonary hypertension resulting in the inability to carry out any physical activity without symptoms. These patients manifest signs of right heart failure. Dyspnea and/or fatigue may be present at rest, and discomfort is increased by any physical activity.

Comments for page:

SIXM:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

Logo	Biomarker/Genotype Sample Collection	{visit.label}
		ID: {ID}

1. Date of collection: BIOM:GCOLLDA / BIOM:GCOLLMO / BIOM:GCOLLYR
   Day Month Year

2. Specimen bar codes:

Description	Bar Code Number	Not Collected
EDTA Whole Blood	BIOM:EDTA	(BIOM:EDTANC)
FTA Blood Spot Card	BIOM:CARD	(BIOM:CARDNC)
Heparin Pellet Sample (2-mL Cryovial)	BIOM:SHPS1	(BIOM:SHPS1NC)
Heparin Pellet Sample (2-mL Cryovial)	BIOM:SHPS2	(BIOM:SHPS2NC)
Heparin Pellet Sample (2-mL Cryovial)	BIOM:SHPS3	(BIOM:SHPS3NC)
Heparin Pellet Sample (2-mL Cryovial)	BIOM:SHPS4	(BIOM:SHPS4NC)
Heparin Pellet Sample (2-mL Cryovial)	BIOM:SHPS5	(BIOM:SHPS5NC)
BNP Sample (2-mL Cryovial)	BIOM:BNP	(BIOM:BNPNC)
Heparin Plasma Sample (2-mL Cryovial)	BIOM:HPS1	(BIOM:HPS1NC)
Heparin Plasma Sample (2-mL Cryovial)	BIOM:HPS2	(BIOM:HPS2NC)
Heparin Plasma Sample (2-mL Cryovial)	BIOM:HPS3	(BIOM:HPS3NC)
Heparin Plasma Sample (2-mL Cryovial)	BIOM:HPS4	(BIOM:HPS4NC)
Citrated Plasma Sample (2-mL Cryovial)	BIOM:CPS1	(BIOM:CPS1NC)
Citrated Plasma Sample (2-mL Cryovial)	BIOM:CPS2	(BIOM:CPS2NC)
Citrated Plasma Sample (2-mL Cryovial)	BIOM:CPS3	(BIOM:CPS3NC)
Citrated Plasma Sample (2-mL Cryovial)	BIOM:CPS4	(BIOM:CPS4NC)
Citrated Plasma Sample (2-mL Cryovial)	BIOM:CPS5	(BIOM:CPS5NC)
Serum Sample (2-mL Cryovial)	BIOM:SS1	(BIOM:SS1NC)
Serum Sample (2-mL Cryovial)	BIOM:SS2	(BIOM:SS2NC)
Serum Sample (2-mL Cryovial)	BIOM:SS3	(BIOM:SS3NC)
Serum Sample (2-mL Cryovial)	BIOM:SS4	(BIOM:SS4NC)

Serum Sample (2-mL Cryovial)	BIOM:SS5	(BIOM:SS5NC)

Comments for page:

BIOM:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

Logo	Subject Disposition	{visit.label}
		ID: {ID}

1. Date of Screening/Observational Follow-Up Informed Consent: SUBD:SCRFUDA / SUBD:SCRFUMO / SUBD:SCRFUYR
   Day Month Year

2. Subject disposition after **Screening:**

- ☐ (SUBD:DISP) Subject enrolled in Observational Follow-Up Study
- ☐ (SUBD:DISP) Subject enrolled in Main Interventional Trial

3. For subjects not enrolled in the **Main Interventional Trial:**

Reason not enrolled in the Main Interventional Trial:

- ☐ (SUBD:REASON) Screening data do not satisfy Main Interventional Trial inclusion/exclusion criteria
- ☐ (SUBD:REASON) Investigator decision, specify:
  SUBD:ID_SP
- ☐ (SUBD:REASON) Subject or parent/guardian decision
- ☐ (SUBD:REASON) Adverse Event, specify:
  SUBD:AE_SP
- ☐ (SUBD:REASON) Lost to follow-up
- ☐ (SUBD:REASON) Other, specify:
  SUBD:OT_SP

Comments for page:

SUBD:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

Logo	Protocol Deviation	{visit.label}
		ID: {ID}

Complete a separate form for <u>each</u> protocol deviation.

1. Date protocol deviation occurred: DEVI:DEVDA (Day) / DEVI:DEVMO (Month) / DEVI:DEVYR (Year)

2. Type of deviation:

- ☐ (DEVI:TYPE) Randomization or masking error
- ☐ (DEVI:TYPE) Dosing error: Did dosing error result in overdose? ☐ (DEVI:OD) No ☐ (DEVI:OD) Yes
- ☐ (DEVI:TYPE) Missed visit
- ☐ (DEVI:TYPE) Missed study procedure/lab test
- ☐ (DEVI:TYPE) Visit out of window: **Early by** DEVI:VISTEA days or **Late by** DEVI:VISTLA days
- ☐ (DEVI:TYPE) Study procedure/lab test out of window: **Early by** DEVI:PROCEA days or **Late by** DEVI:PROCLA days
- ☐ (DEVI:TYPE) Study drug not returned
- ☐ (DEVI:TYPE) Study Drug Diary not returned
- ☐ (DEVI:TYPE) Brief Pain Inventory not returned
- ☐ (DEVI:TYPE) Error in Informed Consent
- ☐ (DEVI:TYPE) Inclusion or exclusion criteria not met, **specify:**

	Check <u>all unmet</u> criteria for <u>this</u> deviation	
	Inclusion Criteria	**Exclusion Criteria**
Screening	☐ (DEVI:INCSC1) 1. Male or female, 12 years of age or older ☐ (DEVI:INCSC2) 2. Diagnosis of sickle cell disease ☐ (DEVI:INCSC3) 3. Provision of informed consent and, where applicable, assent	

Main Interventional Study	☐ (DEVI:INCMS1) 1. Male or female, 12 years of age or older ☐ (DEVI:INCMS2) 2. If female, reliable birth control or not able to bear children ☐ (DEVI:INCMS3) 3. Electrophoretic documentation of sickle cell disease ☐ (DEVI:INCMS4) 4. At least mild pulmonary hypertension ☐ (DEVI:INCMS5) 5. If undergoing right heart catherterization, pulmonary capillary wedge pressure ≤24 mmHg ☐ (DEVI:INCMS6) 6. Six-minute walk distance of 150-500 m ☐ (DEVI:INCMS7) 7. Ability to complete protocol-scheduled assessments during 16-week, double-blind phase ☐ (DEVI:INCMS8) 8. Provision of informed consent and, where applicable, assent	☐ (DEVI:EXCMS1) 1. Current pregnancy or lacation ☐ (DEVI:EXCMS2) 2. Any of: • Stroke within last 6 weeks • New diagnosis of pulmonary embolism within last 3 months • History of retinal detachment/hemorrhage in last 6 months • History of sustained priapism • Non-arteritic anterior ischemic optic neuropathy (NAION) in one or both eyes • Any unstable (chronic or acute) condition that will prevent study completion ☐ (DEVI:EXCMS3) 3. Subject taking nitrate-based vasodilator(s) (including, but not limited to nicorandil [available in the UK only]), prostacyclin (inhaled, subcutaneous or intravenous) or endothelin antagonists. Subjects taking calcium channel blockers will be allowed to participate provided they are on a stable dose for ≥ 3 months. ☐ (DEVI:EXCMS4) 4. Left ventricular ejection fraction < 40% or CS ischemic, valvular or constrictive heart disease ☐ (DEVI:EXCMS5) 5. In other research study with investigational drug except hydroxyurea ☐ (DEVI:EXCMS6) 6. Acute or chronic impairment (other than dyspnea) limiting ability to comply ☐ (DEVI:EXCMS7) 7. Tonsillectomies for sleep apnea within 3 months prior to randomization ☐ (DEVI:EXCMS8) 8. Active therapy for pulmonary hypertension ☐ (DEVI:EXCMS9) 9. Protease inhibitor therapy for HIV treatment ☐ (DEVI:EXCMS10) 10. Potent CYP3A4 inhibitor therapy (e.g., itraconazol, rintonavir, ketoconazole)

Observational Follow-up Study	☐ (DEVI:INCFU1)	1. Satisfaction of screening criteria	
	☐ (DEVI:INCFU2)	2. Ability to maintain follow-up contact	
	☐ (DEVI:INCFU3)	3. Failure to satisfy eligibility requirements of Main Interventional Trial	
	☐ (DEVI:INCFU4)	4. Provision of informed consent and, where applicable, assent	

☐ (DEVI:TYPE) `Other type of deviation,` specify: `DEVI:OTDV_SP`

---

3. Description of deviation and reason it occurred:

`DEVI:DESC`

---

4. Was a protocol waiver granted for the deviation?

☐ (DEVI:WAIVER) `No` ☐ (DEVI:WAIVER) `Yes`

---

5. Study visit at which deviation occurred:

☐ (DEVI:VISIT) `Screening`

☐ (DEVI:VISIT) `Baseline`

☐ (DEVI:VISIT) `Week 6`

☐ (DEVI:VISIT) `Week 10`

☐ (DEVI:VISIT) `Week 16`

☐ (DEVI:VISIT) `Early Termination Visit`

☐ (DEVI:VISIT) `Observational Follow-Up Study`

☐ (DEVI:VISIT) `Open-Label Follow-Up Study`

☐ (DEVI:VISIT) `Not Applicable`

☐ (DEVI:VISIT) `Other,` specify: `DEVI:VISITSP`

6. Steps taken to resolve and prevent recurrence: DEVI:STEPS

7. Did protocol deviation result in an adverse event? ☐ (DEVI:AE) No ☐ (DEVI:AE) Yes

If **Yes,** report on the Adverse Events form.

8. Will the subject continue in the study? ☐ (DEVI:CONT) No ☐ (DEVI:CONT) Yes

9. Did deviation meet reporting requirements for your site's IRB? ☐ (DEVI:IRB) No ☐ (DEVI:IRB) Yes

If **Yes,** date reported to IRB: DEVI:IRBDA / DEVI:IRBMO / DEVI:IRBYR

Day Month Year

Comments for page: DEVI:COMM

Submit Query Cancel Form Completion Help Print Rho

Logo	Adverse Events	{visit.label}
		ID: {ID}

1. Adverse Event/Diagnosis: AEXP:AETEXT

2. AE Start Date: AEXP:STARTDA / AEXP:STARTMO / AEXP:STARTYR
Day Month Year

3. AE Stop Date: AEXP:STOPDA / AEXP:STOPMO / AEXP:STOPYR
Day Month Year

4. Severity: AEXP:SEVRTY — Q4: Death, Life-Threatening, Mild, Moderate, Severe

5. Relationship to sickle cell disease? AEXP:RELSCD

6. Relationship to pulmonary hypertension? AEXP:RELPH

7. Relationship to study drug: AEXP:RELSDRG

8. Relationship to study procedure: AEXP:RELSP

Valid for Q5-Q8:
Definitely Related;
Possibly Related;
Probably not related/remote;
Probably related;
Unrelated

If not **Unrelated,** specify: AEXP:SP_SP

9. Outcome: AEXP:OUTCOME — Q9:Death; Ongoing; Ongoing at end of follow-up; Present at death, not contributing to death; Resolved with sequelae; Resolved without sequelae

10. Action taken with study drug: AEXP:ACTION — Q10: None; Study drug interrupted/modified; Study drug permanently discontinued;

11. Serious? ☐ (AEXP:SERIOUS) No ☐ (AEXP:SERIOUS) Yes

If **Yes,** a) Complete the **Serious Adverse Event** section below and have the Clinical Investigator review and electronically sign the form by clicking the Sign button when prompted.

b) When submitting an initial SAE or a follow-up SAE, please notify Rho Product Safety by either sending an email to rho_productsafety@rhoworld.com or calling the SAE hotline at 888-746-7231.

Has Rho Product Safety been notified? ☐ (AEXP:NTFY) No ☐ (AEXP:NTFY) Yes

Complete this section for a Serious Adverse Event only.

12. Seriousness:

(Check all that apply)

☐ (AEXP:SAE1) Life-threatening

☐ (AEXP:SAE2) Required hospitalization or prolongation of existing hospitalization

☐ (AEXP:SAE3) Congenital anomaly

☐ (AEXP:SAE4) Disabling/incapacitating

☐ (AEXP:SAE5) Important medical event

☐ (AEXP:SAE6) Fatal

If **Fatal:**

a. Date of death: AEXP:DEATHDA / AEXP:DEATHMO / AEXP:DEATHYR
Day Month Year

b. Primary cause of death:

AEXP:CAUSE

c. Was an autopsy performed? ☐ (AEXP:AUTOP) No ☐ (AEXP:AUTOP) Yes

---

13. Possible contributing factors to SAE other than study drug:
(Check all that apply)

☐ (AEXP:FACT1) Underlying disease being studied

☐ (AEXP:FACT2) Treatment failure

☐ (AEXP:FACT3) Concurrent illness, specify: AEXP:FACT3SP

☐ (AEXP:FACT4) Concurrent medication (specify in Number 15 below)

☐ (AEXP:FACT5) Study procedure, specify AEXP:FACT5SP

☐ (AEXP:FACT6) Other, specify AEXP:FACT6SP

---

14. Did subject receive study medication (sildenafil capsules or matching placebo)? ☐ (AEXP:SDYMED) No ☐ (AEXP:SDYMED) Yes

If **Yes:**

a. Phase: ☐ (AEXP:DBFU) Double Blind Phase ☐ (AEXP:DBFU) Open Label Follow-Up Phase

b. Dose: AEXP:DOSE mg TID PO

c. Start Date: AEXP:DSTDA / AEXP:DSTMO / AEXP:DSTYR
Day Month Year

d. Stop Date: AEXP:DSPDA / AEXP:DSPMO / AEXP:DSPYR
Day Month Year

e. Ongoing? ☐ (AEXP:ONGO) No ☐ (AEXP:ONGO) Yes

---

15. Relevant concomitant medications at time of Serious Adverse Event: ☐ (AEXP:CMEDNA) None

☐ (AEXP:CMEDPR) Previously Reported with SAE: AEXP:WITHSAE

		Remove Medication Record			
Name	**Total Daily Dose**	**Units**	**Start Date / Stop Date (Day/Month/Year)**	**Ongoing?**	**Suspect Causal Relationship?**
SAEM:NAME	SAEM:MDOSE	SAEM:MUNITS	SAEM:MSTRTDA / SAEM:MSTRTMO / SAEM:MSTRTYR SAEM:MSTPDA / SAEM:MSTPMO / SAEM:MSTPYR	☐ (SAEM:MONGO) No ☐ (SAEM:MONGO) Yes	☐ (SAEM:RELATE) No ☐ (SAEM:RELATE) Yes

Add Medication Record

---

16. Treatments/procedures for SAE: ☐ (AEXP:TREATNA) None

☐ (AEXP:TREATPR) Previously Reported with SAE: AEXP:TREATSP

	Remove Treatment Record			
Treatment/Procedure	**Total Daily Dose (If Applicable)**	**Units (If Applicable)**	**Start Date / Stop Date (Day/Month/Year)**	**Ongoing?**
SAET:TREAT	SAET:TDOSE	SAET:TUNITS	SAET:TSTRTDA / SAET:TSTRTMO / SAET:TSTRTYR SAET:TSTPDA / SAET:TSTPMO / SAET:TSTPYR	☐ (SAET:TONGO) No ☐ (SAET:TONGO) Yes

Add Treatment Record

17. Relevant medical history (Include only relevant past or concurrent medical disorders, surgeries, etc. that may help explain the SAE): ☐ (AEXP:HISTNA) None

☐ (AEXP:HISTPR) Previously Reported with SAE: AEXP:HISTSP

	Remove History Record	
Condition	**Start Date** **Stop Date** **(Day/Month/Year)**	**Ongoing?**
SAEH:COND	SAEH:HSTRTDA / SAEH:HSTRTMO / SAEH:HSTRTYR SAEH:HSTPDA / SAEH:HSTPMO / SAEH:HSTPYR	☐ (SAEH:HONGO) No ☐ (SAEH:HONGO) Yes

Add History Record

18. Relevant laboratory/diagnostic tests: ☐ (AEXP:LABNA) None

☐ (AEXP:LABPR) Previously Reported with SAE: AEXP:LABSP

	Remove Lab/Test Record	
Lab/Test	**Date** **(Day/Month/Year)**	**Results/Comment**
SAEL:TEST	SAEL:LDATEDA / SAEL:LDATEMO / SAEL:LDATEYR	SAEL:RESULT
	Normal Range (If applicable):	SAEL:RANGE

Add Lab/Test Record

19. Weight: AEXP:WEIGHT ☐ (AEXP:WTUNITS) lb ☐ (AEXP:WTUNITS) kg

20. Height: AEXP:HEIGHT ☐ (AEXP:HTUNITS) in ☐ (AEXP:HTUNITS) cm

21. Narrative/Comments (provide a textual description of the SAE including chronological clinical presentation and evolution of the SAE and associated signs/symptoms):

☐ (AEXP:NARRPR) Previously Reported with SAE: AEXP:NARRSP

Narrative/Comments:

AEXP:NARRATE

Comments for page:

AEXP:COMM

Submit Query | Cancel | Form Completion Help | Print | Rho

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Adverse Events

Adverse Events Form	During **Screening**, adverse events should be reported if they either begin or worsen from the time the subject signs informed consent for Screening/Observational Follow-Up Study through 7 days after the last Screening procedure **and** if they are considered by the investigator to be possibly associated with a study procedure. During the **Main Interventional Trial and Open-Label Follow-Up**, adverse events should be reported if they begin or worsen from the time the subject signs informed consent for Main Interventional Trial through either the last dose of study drug OR until study discontinuation (for those consented subjects who were never treated).
AE/Diagnosis	Enter the diagnostic term for the Adverse Event, if a diagnosis is available.. If a definitive diagnostic term is not available, enter a description of the condition, such as its symptoms, signs, and/or findings. If a definitive diagnosis becomes available at a later time, update the form with that diagnosis.
AE Start Date	Record the date of onset for the AE, providing as complete a date as possible.

AE Stop Date	Record the stop date for each AE, providing as complete a date as possible. If the AE is continuing, leave the Stop Date blank.
Severity	Enter the response that corresponds to the severity of the adverse event, using the following scale: 1. Mild. Awareness of sign, symptom, or event, but easily tolerated; does not interfere with usual daily activities or tasks. 2. Moderate. Discomfort enough to cause interference with usual daily activity; may warrant therapeutic intervention. 3. Severe. Incapacitating; inability to perform usual activities and daily tasks; significantly affects clinical status; requires therapeutic intervention. 4. Life-threatening. Adverse event is life-threatening. 5. Death. Adverse event causes death.
Related to Sickle Cell?	Enter the response that best describes the relationship of the adverse event to sickle cell disease.
Related to Pulmonary Hypertension?	Enter the response that best describes the relationship of the adverse event to pulmonary hypertension.
Relationship to Study Drug	Enter the response that best describes the relationship of the adverse event to use of the study drugl.

Relationship to Study Procedure	Enter the response that best describes the relationship of the adverse event to any study procedure. If Relationship to Study Procedure is any thing except Unrelated, specify the study procedure.
Outcome	Use the drop-down box to select the response that best describes the outcome of the adverse event. If the adverse event is ongoing and the outcome is yet to be determined, leave Outcome blank. The resulting query will serve as a reminder that the AE should be reviewed at the subject's next visit.
Action Taken with Study Drug	Enter the response that best describes what action was taken with the study drug. If the study drug was temporarily or permanently discontinued, there should be a corresponding entry on the Study Drug Dosing form.
Serious?	Indicate whether the adverse event meets the definition of serious by checking No or Yes. If Yes (Adverse Event is Serious): • Complete the Serious Adverse Event (SAE) section of the form. • Have the Clinical Investigator review and electronically sign the form in RhoEDC. • Notify Rho Product Safety of the Serious Adverse Event. If No (AE is not Serious), the Serious Adverse Event (SAE) section of the form should be left blank.

SAE - Seriousness	Check the criteria for "seriousness" met by the SAE. Check all that apply. At least one criteria must be met in order for the AE to be considered an SAE. If the AE was Fatal, provide: • Date of death • Primary cause of death • Whether an autopsy was performed.
SAE - Contributing Factors	Check any factors other than study drug that possibly contributed to the SAE. Check all that apply. If Concurrent Illness, specify the suspected illness. If Study Procedure, specify the suspected procedure. If a possible contributing factor is not listed, check Other and describe the suspected the other contributing factor.
SAE - Study Medication	Check No or Yes to indicate if the subject received study medication (either sildenafil capsules or matching placebo). If Yes, provide: • The phase of the study (either Double Blind Phase or Open Label Follow-Up Phase) • Dose in Mg TID PO • Study drug start date • Study drug end date (if drug not ongoing) • Whether the subject is currently taking study medication

SAE - Relevant Concomitant Medications	Concomitant Medications should be recorded on this form **only** if the investigator considers them to be relevant to the SAE. Check None if there are no concomitant medications relevant to the SAE. If there are relevant concomitant medications, provide for each one: • Medication name • Total daily dose • Dosage units • Start date and stop date • Whether the medication is ongoing • Whether a causal relationship between the medication and the SAE is suspected Use the Add Medication Record button to create a row for each relevant medication. Use the Previously Reported checkbox if this information was reported with a previous SAE report. If so, identify the previous SAE.
SAE - Treatments/ Procedures	Check None if no treatments or procedures were prescribed with this SAE. If there were treatment or procedures to report for this SAE, provide for each one: • Name of treatment or procedure • Total daily dose (if applicable) • Units (if applicable) • Start date and stop date • Whether the treatment/procedure is ongoing Use the Add Treatment Record button to create a row for each treatment or procedure.

	Use the Previously Reported checkbox if this information was reported with a previous SAE report. If so, identify the previous SAE.
SAE - Relevant Medical History	Medical History should be recorded on this form **only** if the investigator considers it to be relevant to the SAE. Check None if there is no relevant medical history that would help explain the SAE. Use the Add History button to create a row for each relevant medical history item. Provide for each one: • The medical condition • Start date and stop date. • Whether the condition is ongoing. Use the Previously Reported checkbox if this information was reported with a previous SAE report. If so, identify the previous SAE.
SAE - Relevant Laboratory/ Diagnostic Tests	Laboratory and/or diagnostic tests should be recorded on this form **only** if the investigator considers them to be relevant to the SAE. Check None if there are no relevant tests to report with this SAE. Use the Add Lab/Test Record to create a row for each relevant test. Provide for each: • Name of the test • Result/lab value • Units for the result/lab value • Normal range for the test Use the Previously Reported checkbox if this information was reported with a previous SAE report. If so, identify the previous SAE.

SAE - Weight and Height	Record the subject's weight and height at the time of the SAE and the appropriate unit for each.
SAE - Narrative/ Comments	Record the narrative description of the SAE, including chronological clinical presentation and evolution of the SAE and associated signs/symptoms. Use the Previously Reported checkbox if this information was reported with a previous SAE report. If so, identify the previous SAE.
Comments for page	Record any pertinent comments for this page only.
See also Chapter 15 of the MOO	

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Biomarker/Genotype Sample Collection

Date of Collection	Record the date the samples were collected. **Note**: If no sample was collected, provide an explanation in the Comments field at the bottom of the form.
Is this the Screening Visit?	Answer No or Yes.
Specimen Bar Code	Record the specimen bar code number from each sample collected.
Comments for page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Chemistry

Date of Collection	Record the date the sample was collected for analysis.
Lab Value	Record the result for each test.
Unit	Confirm that the unit reported on the lab slip is the same as the unit on the form. If the unit on the lab slip is unequal to a unit on the form, contact the coordinating center. For help with SI conversions: http://nephron.com/cgi-bin/SI.cgi AND http://www.unc.edu/~rowlett/units/scales/clinical_data.html **Note:** The following units are equivalent:
	10^3cells/µL, 1000/µL, th/µL, K/µL, 10^3/mm^3, 1000/mm^3, th/mm^3, K/mm^3, and 10^9/L
	10^6/µL, mil/µL, M/µL, 10^6/mm^3, mil/mm^3, M/mm^3, 10^{12}/L, and mil/mcL

Clinical Significance	Check the one box that best describes the clinical significance of each result. Clinical significance should be determined by the investigator based on his or her judgment and knowledge of the individual subject and the study population. Generally, a result is deemed "clinically significant" if it requires treatment, re-testing or other follow-up. If the test result is judged clinically significant **and** a new Adverse Event, complete an Adverse Event form.
Comments for page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Demographics

Demographics Form	The demographics form should be completed for each subject at the time of his/her initial screening. If the subject does not meet the criteria to be enrolled in the Main Interventional Trial at his/her initial screening visit but is subsequently **re-screened and enrolled** in the MIT, the information on the form should be updated.
Date of Assessment	Record the date on which the subject's demographic information was collected.
Date of Birth	Enter the subject's date of birth.
Gender	Enter the subject's gender at birth.
Ethnicity	Ethnicity should be determined by the subject. Check the one box that corresponds to the subject's assessment of his/her ethnicity as it relates to the Hispanic or Latino population.

Race	Race should be determined by the subject. Check the box or boxes that correspond to the subject's assessment of his/her race. Check all that apply. Subjects may self-identify themselves as belonging to multiple race categories. If the subject self-identifies his/her race as Black, the subject should characterize further if possible. If the subject does not further characterize his/her race, enter Not Otherwise Specified under Black. If the subject self-identifies his/her race as Unknown, check Unknown. If the subject declines to identify his/her race, check No Response. If the subject's self-identified race is not listed, check Other and specify the subject's race as self-identified in the space provided. **Note:** Subjects can decline to provide information about Ethnicity and Race although efforts should be made to collect this information. Declining to present this information should not affect subject enrollment or randomization.
Countries of Ancestry	Enter countries of ancestry as self-identified by the subject. If the subject indicates that his/her countries of ancestry are unknown, check Unknown.
Medical Care Received Outside the U. S. or U.K.	Check No or Yes to indicate whether the subject has received medical care outside the United States or United Kingdom for period(s) exceeding 1 year. If Yes, enter the number of years the subject received medical care outside the U.S. or U.K.

Where was Subject Born?	Enter where the town, city or state and the country where the subject was born.
Demographic Information on Subject's Mother and Father	Enter demographic information regarding birthplace, ethnicity and race for the subject's mother and father, using the instructions above.
Parents Living ?	Check No or Yes to indicate whether the subject's mother and father are alive. If No, specify the parent's cause of death if known. If the cause of death is unknown, enter 'Unknown.'
Family Diseases and Disorders	List any diseases and/or disorders known to be present on the mother's side of the family and on the father's side of the family in their respective columns. If unknown, enter 'Unknown.'
Number of Full Siblings	Enter the number of full siblings the subject has, or check Unknown the number is unknown. If the subject has no full siblings, enter '0'.
Number of Half Siblings	Enter the number of half siblings the subject has, or check Unknown the number is unknown. If the subject has no half siblings, enter '0'.

Sibling Records	Click the Add a Sibling Record button for each of the subject's full and half siblings and report the following information for each: • Relationship to subject (full or half sibling) • Year of sibling's birth • Whether the sibling has sickle cell trait • Whether the sibling has sickle cell disease • Whether the sibling is living • If the sibling is deceased, year of death and cause of death Use the Remove button to delete any sibling records that are created in error. Use caution in deleting records, as deleted records cannot be recovered after the form is updated.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Protocol Deviation

Protocol Deviation Form	The Protocol Deviation form should be completed to document any departure from the study protocol. Complete a separate form for each deviation.
Protocol Deviation Date	Record the date the protocol deviation occurred.
Type of Deviation	Check the one box that best describes the protocol deviation being reported. **Dosing error:** If the deviation was a dosing error, check No or Yes to indicate whether the dosing error resulted in an overdose. **Visit out of window:** If the deviation was a study visit occurring outside of the protocol window, enter the number of days by which the visit was either early or late. **Procedure/test out of window:** If the deviation was a procedure or testing occurring outside of the protocol window, enter the number of days by which it was either early or late. **Inclusion/Exclusion criteria not met:** If the deviation was that protocol inclusion or exclusion criteria were not met, indicate which criteria were not met. Check all that apply. **Other:** If the protocol deviation is of a type not listed, check Other and specify in the space provided.

Deviation Description	Enter a description of the deviation and the reason it occurred.
Protocol Waiver Granted?	Check No or Yes to record whether a protocol waiver was granted.
Study Visit at Which Deviation Occurred	Check the one box to indicate at which study visit the protocol deviation occurred. If the deviation did not occur at a study visit, check Other and specify in the space provided.
Steps Taken to Resolve and Prevent Recurrence	Describe any steps that were taken to resolve the deviation and to prevent it from occurring again.
Adverse Event?	Check No or Yes to indicate whether the protocol deviation resulted in an Adverse Event. If the deviation occurred while the subject was enrolled in the Observational Follow-Up Study, which does not include monitoring for Adverse Events, check Not Applicable. **Note:** If the deviation resulted in an Adverse Event, be sure to report it on an Adverse Events form.
Will Subject Continue in Study?	Check No or Yes to indicate whether the subject will continue in the study despite the deviation. If No, be sure to complete the Study Completion/Early Termination form.

Reporting Deviation to IRB?	Check No or Yes to indicate whether the deviation meets the reporting requirement for your site's IRB. If Yes, record the date the deviation was reported to the IRB.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Echocardiogram (Local)

Date of Procedure	Record the date the echocardiogram was performed.
Last Study Drug Dose	Record the date and time of the subject's last dose of study drug before the echo procedure. (At Screening, this question is not applicable and does not appear on the form.) **Note**: The echocardiogram should be performed when the subject is on peak dose of study drug dose, which is be 1-1.5 hours after taking the medication.
Time of Procedure	Record the time the echocardiogram was performed, using 24-hour clock. **Note**: The echocardiogram should be performed when the subject is on peak dose of study drug dose, which is be 1-1.5 hours after taking the medication.
Blood Pressure	Record subject's blood pressure at the time of the procedure.

Tricuspid Regurgitant Jet Velocity	Enter the Tricuspid Regurgitant Jet Velocity (TRV) recorded by the echo. If the TRV could not be determined, check Not Detectable. **Note:** Not Detectable is not an acceptable response at Screening and Baseline. In order to be enrolled in the Main Interventional Trial, the subject must have a TRV ≥ 2.7 m/s.
Right Atrial Pressure	Check the box that corresponds to the subject's estimated right atrial pressure in mmHg.
LV Function	Check Normal or Abnormal.
LV Ejection Fraction	Enter the subject's LV ejection fraction.
Regurgitation Findings	Enter aortic, mitral and tricuspid regurgitation findings by checking None, Trace, Mild, Mild-Moderate, Moderate, Moderate-Severe or Severe for each.
Other Significant Findings	Enter any other relevant findings from the echocardiogram.
Bar Code	Enter the bar code(s) from the label used to track the shipment of the echo recording. The bar code may be entered by typing in the number or by scanning the bar code with a hand-held scanner. If scanning, check to see that the cursor is in the Bar Code field before scanning.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Hematology

Date of Collection	Record the date the sample was collected for analysis.
Lab Value	Record the result for each test.
Unit	Confirm that the unit reported on the lab slip is the same as the unit on the form. If the unit on the lab slip is unequal to a unit on the form, contact the coordinating center. Note: Formula for MCHC is Hb x 100/Hct, if not on print out from lab. For help with conversions: http://nephron.com/cgi-bin/SI.cgi AND http://www.unc.edu/~rowlett/units/scales/clinical_data.html **Note:** The following units are equivalent:
	10^3cells/µL, 1000/µL, th/µL, K/µL, 10^3/mm^3, 1000/mm^3, th/mm^3, K/mm^3, and 10^9/L
	10^6/µL, mil/µL, M/µL, 10^6/mm^3, mil/mm^3, M/mm^3, 10^{12}/L, and mil/mcL

Clinical Significance	Check the one box that best describes the clinical significance of each result. Clinical significance should be determined by the investigator based on his or her judgment and knowledge of the individual subject and the study population. Generally, a result is deemed "clinically significant" if it requires treatment, re-testing or other follow-up. If the test result is judged clinically significant **and** a new Adverse Event, complete an Adverse Event form.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

HIV Test

Date of Collection	Record date the sample was collected for HIV testing.
Result	Check Negative or Positive. If the test was Positive, check No or Yes to indicate whether the subject is on protease inhibitor therapy for treatment of HIV.
Comments for page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Medical History
Part 1: Diagnosis, Transfusion, Reproductive & Social Histories

Medical History Form	For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history should be collected via a review of the subject's medical records, if available. If the subject does not meet the criteria to be enrolled in the Main Interventional Trial at his/her initial screening visit but is subsequently **re-screened and enrolled** in the MIT, the information on the form should be updated.
Date of Assessment	Record the date the subject's medical history was assessed for the purposes of the study. **Note**: If the subject was not randomized in the Main Interventional Trial after an initial screening but rather was subsequently rescreened and randomized, the subject's medical history should be updated to reflect his/her status at the time of the re-screening. The Date of Assessment in that case would be that of the subsequent rescreening that resulted in randomization.

Study Diagnosis History	Check the box that best describes the subject's sickle cell genotype. If Other, specify the genotype. Record the date the subject was diagnosed with pulmonary hypertension. Provide as complete a date as possible. If a complete date is not known, enter as much of the date as can be determined and leave blank any parts that are unknown. **Note**: Blank date parts will cause an automatic query. If the date part is blank because a complete date is unknown and is impossible to determine, override the automatic query. Provide an appropriate override reason, such as "Day Unknown" or "Month Unknown."
Transfusion History	Check the box that best describes the number of transfusions the subject has had in his/her lifetime. Check No or Yes to indicate whether the subject is on chronic transfusion therapy. If Yes, enter the date chronic transfusion therapy started. If a complete date is not known, enter as much of the date as can be determined and leave blank any parts that are unknown. **Note**: Blank date parts will cause an automatic query. If the date part is blank because a complete date is unknown and is impossible to determine, override the automatic query. Provide an appropriate override reason, such as "Start Day Unknown" or "Month Unknown." If subject has had transfusion, enter the following: • Date of last transfusion • The type of transfusion given, Simple, Exchange or Other. • Number of units transfused. Also, record whether the subject has had previous transfusion reactions. If the subject has not had previous transfusion reactions, check None. If the

	subject has had reaction, check the box or boxes that best describe the reaction. If Other, specify the reaction.
Reproductive History, Female	The reproductive history section of the form applies only to female subjects. If subject is male, check Not Applicable. If the subject is female, check the box that best describes the subject's reproductive status: Pre-menarche, Post-menarche but pre-menopausal, or Post-menopausal . If subject is Post-menarche but pre-menopausal, enter the following: • Age of menarche • Menstual cycle length in days • Whether menstual cycle is regular If subject is Post-menopausal, enter the following: • Age of menarche • Age at onset of menopause • Month and Year of last menstrual period **Note:** For the subject to be considered post-menopausal, at least one year must have passed since the last menstual period. Also enter the number of pregnancies and live births the subject has had. Enter 0 if the subject is pre-menarche or otherwise has never been pregnant or has never given birth.

Social History - Smoking	Check the box that best describes subject's smoking history: None, Current Smoker, or Former Smoker. If current or former smoker, enter the following: • Year subject started smoking • Maximum packs smoked per day If the subject is a former smoker, also enter the year subject stopped smoking
Social History - Alcohol	Check the box that best describes subject's history of alcohol use: None, Current Alcohol Use, or Former Alcohol Use. If subject currently drinks or formerly drank alcohol, enter the maximum number of drinks consumed per week.
Social History - Drug Use	Check the box that best describes subject's history of using recreational or illegal drugs: None, Current Drug Use, or Former Drug Use. If subject currently uses or formerly used drugs, check the boxes that correspond to the drugs used. Check all that apply. If Other, specify the drug used.
Data Collection	Check the box that best describes how the information on this form was collected: • All or most per subject (or parent/guardian) report; not confirmed via medical record • All or most confirmed via medical record • Other If the best choice is Other, describe how the information was collected.

	Note: For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history is expected to be collected via a review of the subject's medical records, if available.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Medical History
Part 2: Surgical & Disease Histories

Medical History Form	For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history should be collected via a review of the subject's medical records, if available. If the subject does not meet the criteria to be enrolled in the Main Interventional Trial at his/her initial screening visit but is subsequently **re-screened and enrolled** in the MIT, the information on the form should be updated.
Date of Assessment	Record the date the subject's medical history was assessed for the purposes of the study. **Note**: If the subject was not randomized in the Main Interventional Trial after an initial screening but rather was subsequently rescreened and randomized, the subject's medical history should be updated to reflect his/her status at the time of the re-screening. The Date of Assessment in that case would be that of the subsequent rescreening that resulted in randomization.

Surgical History	Check No, Yes or Unknown to indicate whether the subject has had any of the listed procedures. For any procedure where the response is Yes, enter the Year Performed. Use the corresponding Comment/ Complications field to record any relevant comments about the procedure, including any complications. If the subject had the same surgery more than once, record the year of the most recent procedure and provide details of prior occurrences in the corresponding Comment/Complications field. If a procedure from the subject's history is not listed, check Yes for Other and click the Add Surgery Record button as needed in order to enter details of each procedure.
Diseases/ Disorders/ Ailments History	For each Disease/Disorder/Ailment category, check No or Yes to indicate whether the subject has history in that category. If Yes (subject has history within the category), check the Yes box beside the specific condition the subject has experienced. Check all that apply within each category. Use Other if the subject's specific ailment is not listed. If the subject reports having history within a category but is unable to identify the specific condition, check 'Subject unsure what problem is/was.'

Diagnostic Tests	Check No or Yes to indicate whether the subject has ever had any of the listed diagnostic tests (MRI, head, MRA, head, Transcranial Doppler (TCD), Echocardiogram, Pulmonary Function Testing, or EKG). If Yes, click the Add Test Record to report the date and result of each test performed. Use the corresponding Comment field to enter any relevant findings or comments from the diagnostic testing. Use the Remove button to delete any test records that are created in error. Use caution in deleting records, as deleted records cannot be recovered after the form is updated.
Data Collection	Check the box that best describes how the information on this form was collected: • All or most per subject (or parent/guardian) report; not confirmed via medical record • All or most confirmed via medical record • Other If the best choice is Other, describe how the information was collected. **Note:** For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history is expected to be collected via a review of the subject's medical records, if available.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Medical History
Part 3: Medications and Pain Histories

Medical History Form	For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history should be collected via a review of the subject's medical records, if available. If the subject does not meet the criteria to be enrolled in the Main Interventional Trial at his/her initial screening visit but is subsequently **re-screened and enrolled** in the MIT, the information on the form should be updated.
Date of Assessment	Record the date the subject's medical history was assessed for the purposes of the study. **Note**: If the subject was not randomized in the Main Interventional Trial after an initial screening but rather was subsequently rescreened and randomized, the subject's medical history should be updated to reflect his/her status at the time of the re-screening. The Date of Assessment in that case would be that of the subsequent rescreening that resulted in randomization.

Medications History	For each class of medications listed, check the box that best describes subject's current and past usage of that group of medications. Check either Currently Using, Used in the Past, or both. For the three groups of pain medications listed (Narcotics, NSAIDs and Other), indicate use only if the subject has taken a medication within the class daily for 30+ days. Use of pain medications for terms shorter than 30+ days should not be recorded on this form. • As applicable, list the specific medications used by the subject within a group of medications. For example, if the subject has used or is using anticonvulsants, list the names of the specific anticonvulsants. • As applicable, check the box that best corresponds to the subject's cumulative lifetime use of a class of medications. For example, if the subject has used medications intermittently for 1-5 years, then mark the box 1-5 years. As applicable, check the box that best describes subject's current and past usage of the specific medications listed within a group. For example, check Currently Using, Used in the Past, or both for each of the narcotics listed by name.
Previous Medication Reactions	Record whether the subject has had previous medication reactions. If the subject has not had previous reactions, check None. If the subject has had reaction, check the box or boxes that best describe the reaction. If Other, specify the reaction. Also, list the specific medication(s) that caused a reaction.

Sickle Cell Pain History - Acute Pain	**Note:** The first section of Sickle Cell Pain History applies to **acute** pain. Enter the following information as regards acute sickle cell pain: • Location: Check the box or boxes that best describe the location of the pain. If Other, specify the location. • Typical pain rating on a 1-10 scale. • Description of quality or type of pain. • Treatment: Check the box or boxes that correspond to how the subject typically treats acute sickle-cell pain. Enter the number of pain crises/pain events experienced by the subject in the last week, dividing the number into the four categories of severity: Mild, Moderate, Severe, Extremely Severe. Enter 0 if the subject has experienced no events of a listed severity. Enter the number of pain crises/pain events experienced by the subject in the last week, month and year. Divide the total number among the four categories of severity: Mild, Moderate, Severe, Extremely Severe. Enter 0 if the subject has experienced no events of a listed severity in the specified time period.
Sickle Cell Pain History - Chronic Pain	Check No or Yes to indicate whether the subject also reports having chronic pain. If Yes, enter the following information as regards chronic sickle cell pain: • Location: Check the box or boxes that best describe the location of the pain. If Other, specify the location. • Typical pain rating on a 1-10 scale. • Description of quality or type of pain. • Treatment: Check the box or boxes that correspond to how the subject typically treats chronic sickle-cell pain.

Data Collection	Check the box that best describes how the information on this form was collected: • All or most per subject (or parent/guardian) report; not confirmed via medical record • All or most confirmed via medical record • Other If the best choice is Other, describe how the information was collected. **Note:** For the Observational Follow-Up Study, the subject's medical history can be collected based on subject self-report. For subjects randomized in the Main Interventional Trial, the medical history is expected to be collected via a review of the subject's medical records, if available.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Physical Examination

Date of Assessment	Record the date of the examination.
Vital Signs	Record the subject's vital signs: • Temperature in degrees Celsius. • Heart rate in beats/minute. • Oxygen saturation and whether the reading was taken with the subject on room air or supplemental oxygen. If the subject was using supplemental oxygen, enter the oxygen flow rate. • Respiratory rate in breaths/minute. • Sitting blood pressure. • Weight in kilograms • Height in centimeters • Body surface area in square meters, using the Mosteller formula. For help with BSA or metric conversions: http://www.halls.md/body-surface-area/bsa.htm AND http://www.teaching-english-in-japan.net/conversion/feet_inches

Body System Categories	Assess the subject's physical condition as it relates to each of the listed body systems. Answer the specific questions related to each body system by checking the boxes or boxes that correspond to the subject's condition. Enter any other findings or comments for each body system in the corresponding Comment/Other findings or abnormalities box.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Pregnancy Test

Date of Assessment	Record the date the pregnancy test was performed. If the pregnancy test was not performed, the date may be left blank.
Pregnancy Test Result	Check Negative, Positive, or Not Done. If Not Done, check the box that best describes why the test was not performed. If the reason is not listed, check Other and specify in the space provided.
Type of Test	Enter the type of pregnancy test used, either Urine or Serum. If a serum pregnancy test was performed, record the HCG result of the test. **Note:** The following units are equivalent: mIU/ML and IU/L
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Randomization/Subject Disposition

Randomization/ Subject Disposition Form	The Randomization/Subject Disposition form should be completed after the subject has completed the Baseline visit in the Main Interventional Trial.
Date of MIT Informed Consent	Enter the date the subject signed informed consent for the the Main Interventional Trial.
Subject Disposition after Baseline	Indicate the subject's status as regards randomization into the Main Interventional Trial after the Baseline visit. Check either: • Subject Randomized in Main Interventional Trial • Subject Not Randomized In Main Interventional Trial/Subject Enrolled In Observational Follow-Up Study
Date Randomized and Randomization Stratum	If the subject was randomized, enter the date of randomization and check the TRV Statrum for the subject.

Reason Not Randomized	If the subject was not randomized, check the reason why not. If the reason was Adverse Event, Investigator Decision or Other, specify the details.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Right Heart Catheterization

Subject's TRV for Enrollment in MIT	Check No or Yes to indicate whether the subject's tricuspid regurgitant jet velocity for enrollment in the Main Interventional Trial was ≥ 3.0 m/s. If Yes, the results of the required right heart catherization procedure should be recorded on this form. If No (subject's TRV < 3.0 m/s), the right heart catherization procedure was not required and the rest of this form should be left blank.
Study Visit	Check the box that corresponds to the study visit for which the right heart cath was performed, either Baseline or Week 16/Early Termination.
Last Dose of Study Drug	If this is the subject's Week 16 or Early Termination visit, record the date and time of the subject's last dose of study drug. Use a 24-hour clock. If this is the subject's Baseline visit, the question is not applicable and should be left blank.
Date of Procedure	Record the date the right heart catherization was performed.

Catheterization Side	Check Left or Right to indicate whether the procedure was performed on the right or left side of the heart. If a left heart catherization was performed, enter an explanation in the comment field at the bottom of the form.
Steps 1-5	Steps 1 through 5 should be completed at the Baseline visit. Step 1, 4 and 5 should be completed at the Week 16 or Early Termination visit. Leave Step 2 and Step 3 blank at the Week 16 or Early Termination visit. For each step performed, record the values corresponding to each of the parameters listed. See protocol section XXX or Chapter XX of the Manual of Operations for details.
Bar Code	If the right heart cath recording was shipped, enter the bar code from the label used to track the shipment. If the recording was not shipped, check 'Recording not sent.' The bar code may be entered by typing in the number or by scanning the bar code with a hand-held scanner. If scanning, check to see that the cursor is in the Bar Code field before scanning.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

6-Minute Walk Test

6-Minute Walk Test Form	The 6-minute walk is the Main Interventional Trial's primary endpoint and must be performed as outlined in Chapter XX of the Manual of Operations. **Language about repeat walk at baseline if not within 15% of Screening.**
Date of Assessment	Record the date the 6-minute walk was performed.
Date/ Time of Last Dose of Study Drug	At visits other than Screening and Baseline, enter the date and time of the subject's last dose of study drug. At Screening and Baseline visits, check Not Applicable for date and time of last study drug dose.
Before Walk	Enter the following from before the start of the walk test: 1. Subject's blood pressure. 2. Subject's heart rate. 3. Subject's oxygen saturation and whether the reading was taken with the subject on room air or supplemental oxygen. If the subject was using supplemental oxygen, enter the oxygen flow rate.

	4. The time the walk started, using a 24-hour clock.
After Walk	Enter the following from after the end of the walk test: 5. Subject's blood pressure. 6. Subject's heart rate. 7. Subject's oxygen saturation and whether the reading was taken with the subject on room air or supplemental oxygen. If the subject was using supplemental oxygen, enter the oxygen flow rate. 8. Distance walked, in meters. 9. Whether the subject stopped before the 6-minute time limit was over. 10. Whether the subject used oxygen during the walk test. If Yes, enter the flow rate and whether the subject carried or pushed the oxygen device during the test. 11. Use the drop-down box to select the correct value for the Borg dyspnea score. 12. Use the drop-down box to select the correct value for the NYHA/WHO classification.
11. Borg Dyspnea Score	Select the value that corresponds to the subject's Borg dyspnea score at the end of the walk.
12. NYHA/ WHO Classification	Check the box that corresponds to the subject's NYHA/ WHO classification at the end of the walk.

Comments for page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Subject Disposition

Date of Screening/ Observational Follow-Up Informed Consent	Enter the date the subject signed informed consent.
Subject Disposition after Screening	Indicate the subject's status as regards enrollment into the Main Interventional Trial or in the Observational Follow-Up Study. Check either: • Subject enrolled in Observational Follow-Up Study • Subject enrolled in Main Interventional Trial
Reason Not Randomized	If the subject was not enrolled in Main Interventional Trial , check the reason why not. If the reason was Adverse Event, Investigator Decision or Other, specify the details.
Comments for Page	Record any pertinent comments for this page only.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Subject Enrollment

Subject ID	Enter the Subject ID in the space provided.

EDC COMPLETION GUIDELINES

walk-PHaSST Protocol

Urinalysis

Date of Collection	Record the urine sample was collected for urinalysis.
Urine Dipstick Chemical Analysis	Check No, Yes or Not Applicable (male subject) to indicate whether the subject is menstruating. Enter the pH value. Enter the Specific Gravity value. Enter the dipstick result for glucose, protein and blood by checking Negative or Positive. If any result is Positive, use the drop-down box to select the code for the result and/or enter the positive value (depending on how the result is reported by the lab). For help with conversions: http://nephron.com/cgi-bin/SI.cgi AND http://www.unc.edu/~rowlett/units/scales/clinical_data.html
Microscopic Exam	Check No or Yes to indicate whether a microscopic exam was performed. If Yes, use the drop-down boxes to enter the value that corresponds to the lab's findings for RBC and WBC. Check No or Yes box for whether there were other abnormal findings on the microscopic exam. If Yes, provide a description of the other abnormal findings.

Overall Assessment of Urinalysis	Check Normal or Abnormal to report the overall assessment of the urinalysis. If Abnormal, check No or Yes to indicate whether the finding represents a new Adverse Event. If Yes, report the AE on the Adverse Event form.
Albumin/ Creatinine Ratio	Record the date the sample was collected for the albumin/creatinine ratio test. Enter the albumin and creatinine values, or enter the ratio itself (depending on how the result is reported by the lab).
Comments for page	Record any pertinent comments for this page only.