### Sickle Cell Anemia Pre-HSCT Data

**Registry Use Only**

<table>
<thead>
<tr>
<th>Sequence Number:</th>
<th>Date Received:</th>
</tr>
</thead>
</table>

This form must be accompanied by Form 2000 – Recipient Baseline Data. All information in the box above, including the date, should be identical with the corresponding Form 2000. Information should come from an actual examination by the Transplant Center physician, or the physician who is following the recipient pre-HSCT, or abstraction of the recipient’s medical records.

If this is a report of a second or subsequent transplant, check here ☐ and continue with question 92.

1. What was the date of diagnosis of Sickle Cell Anemia? [ ]

<table>
<thead>
<tr>
<th>Month</th>
<th>Day</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

2. Was the recipient diagnosed with sickle cell disease at birth (i.e., newborn screening)?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

3. What is the recipient’s sickle cell disease genotype?
   1 ☐ Hb SS
   2 ☐ Hb S beta0 thalassemia
   3 ☐ Hb SC
   4 ☐ Hb S beta+ thalassemia
   5 ☐ other genotype

4. Specify other genotype: ____________________________

5. Did the recipient receive red blood cell transfusions at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

6. Date of first transfusion: [ ]

<table>
<thead>
<tr>
<th>Month</th>
<th>Day</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

   ☐ date unknown

7. Specify the total number of transfusions received prior to the preparative regimen:
   1 ☐ < 5
   2 ☐ 5–10
   3 ☐ > 10

8. Did the transfusion(s) induce red cell alloimmunization?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

9. Specify the number of alloantibodies detected:
   1 ☐ 1
   2 ☐ ≥ 2
   3 ☐ unknown
30. Was iron chelation therapy performed at any time prior to the preparative regimen?

1. Yes
2. No
3. Unknown

31. Date chelation therapy started: [□] Month [□] Day [□] Year [□] Date unknown

32. Specify the predominant route of administration:
1. Intramuscular
2. Intravenous
3. Oral
4. Subcutaneous
5. Other route
6. Unknown

33. Specify other route: __________________________

27. Specify: ______

29. Specify the number of autoantibodies detected:
1. 1
2. ≥ 2
3. Unknown

28. Are red cell autoantibodies present?

1. Yes
2. No
3. Unknown

26. Unknown other

10. Specify the blood group(s) the recipient has developed alloantibodies to:

10.1 Yes
2. No
3. Unknown

Duffy –Fy^a
Kell –K
Kell –k
Kidd –Jk^a
Kidd –Jk^b
Lewis –Le^a
Lewis –Le^b
MNSs –M
MNSs –N
MNSs –S
MNSs –s
Rh –C
Rh –D
Rh –E
Rh –e
Rh –hr^a

22. Unknown Rh –D
23. Unknown Rh –E
24. Unknown Rh –e
25. Unknown Rh –hr^a
26. Unknown other

21. Unknown Rh –C

11. Duffy –Fy^a

12. Kell –K

13. Kell –k

14. Kidd –Jk^a

15. Kidd –Jk^b

16. Lewis –Le^a

17. Lewis –Le^b

18. MNSs –M

19. MNSs –N

20. MNSs –S

21. MNSs –s

22. Rh –C

23. Rh –D

24. Rh –E

25. Rh –e

26. Rh –hr^a

27. Specify: ______

28. Are red cell autoantibodies present?

1. Yes
2. No
3. Unknown

29. Specify the number of autoantibodies detected:
1. 1
2. ≥ 2
3. Unknown

30. Was iron chelation therapy performed at any time prior to the preparative regimen?

1. Yes
2. No
3. Unknown

31. Date chelation therapy started: [□] Month [□] Day [□] Year [□] Date unknown

32. Specify the predominant route of administration:
1. Intramuscular
2. Intravenous
3. Oral
4. Subcutaneous
5. Other route
6. Unknown

33. Specify other route: __________________________
34. Was a liver biopsy performed at any time prior to the preparative regimen?

1: yes
2: no
3: unknown

35. Date of most recent liver biopsy:

- Month
- Day
- Year
- date unknown

36. Was hepatitis present?

1: yes
2: no
3: unknown

37. Specify the severity of hepatitis:

1: mild
2: moderate
3: severe
4: unknown

38. Was siderosis present?

1: yes
2: no
3: unknown

39. Specify the severity of siderosis:

1: mild
2: moderate
3: severe
4: unknown

40. Was fibrosis present?

1: yes
2: no
3: unknown

41. Specify the severity of fibrosis:

1: mild
2: moderate
3: severe
4: unknown

42. Were serial liver biopsies performed?

1: yes
2: no

43. Did the liver biopsies show progressive disease?

1: yes
2: no
3: unknown

44. What was the hepatic iron concentration (HIC)?

1: known
2: unknown

45. Specify HIC: mg/g

46. Is a copy of the biopsy report attached?

1: yes
2: no

47. Were pulmonary function tests (PFTs) performed at any time prior to the preparative regimen?

1: yes
2: no
3: unknown

48. Specify PFT results: (see definitions on the following page)

1: normal
2: Stage 1 disease
3: Stage 2 disease
4: Stage 3 disease
5: Stage 4 disease
6: unknown

49. Is a copy of the PFT report attached?

1: yes
2: no
## Sickle Chronic Lung Disease Staging Criteria (Clinical)

<table>
<thead>
<tr>
<th>Markers</th>
<th>Stage 1</th>
<th>Stage 2</th>
<th>Stage 3</th>
<th>Stage 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest Pain</td>
<td>Recurrent substernal pain and chronic cough</td>
<td>Increased pain over Stage 1</td>
<td>Severe midline crushing chest pain</td>
<td>Severe and prolonged pain with dyspnea at rest</td>
</tr>
<tr>
<td>Blood Gasses</td>
<td>Normal oxygen saturation</td>
<td>Normal oxygen saturation</td>
<td>Hypoxia with partial pressure oxygen (70 mm Hg) during stable periods</td>
<td>Partial pressure oxygen (60 mm Hg) during stable periods</td>
</tr>
<tr>
<td>X-Ray</td>
<td>Decreased distal pulmonary vascularity, hyperexpansion, evidence suggestive of increased interstitial markings</td>
<td>Diffuse, fine interstitial fibrosis</td>
<td>Pulmonary fibrosis</td>
<td>Severe pulmonary fibrosis</td>
</tr>
<tr>
<td>Pulmonary Function Tests *</td>
<td>Decreased FVC, TLC, FEV₁ and FEV₁ / FVC ratio (mild, 80% of predicted normal, or 1 standard deviation below normal)</td>
<td>Decreased FVC, FEV₁, TLC, DCO and FEV₁ / FVC ratio (moderate, 60% of predicted, or 2 standard deviations below normal)</td>
<td>Decreased FVC, REV₁, TLC, DCO and FEV₁ / FVC ratio (severe, 40% of predicted, or 3 standard deviations below normal)</td>
<td>Patient frequently unable to complete testing due to degree of hypoxia</td>
</tr>
<tr>
<td>ECG and ECHO</td>
<td>Left ventricular preponderance persists</td>
<td>Balanced ventricular hypertrophy</td>
<td>Right ventricular hypertrophy and right atrial enlargement. Progressive increase in heart size</td>
<td>Severe right ventricular and right atrial hypertrophy. Ischemic T waves in V1 and V2, and P pulmonale</td>
</tr>
<tr>
<td>Pulmonary Artery Pressure</td>
<td>Normal</td>
<td>Normal</td>
<td>Borderline elevation or normal</td>
<td>Markedly elevated with pulmonary hypertension</td>
</tr>
</tbody>
</table>

* These measurements are based on common methods for comparison of reference values. Abbreviations: FVC = forced vital capacity, TLC = total lung capacity, REV₁ = forced expiratory flow rate

### Specify the sickle cell disease symptoms experienced at any time prior to the preparative regimen:

50. Acute chest syndrome

- 1 □ yes
- 2 □ no
- 3 □ unknown

51. Total number of episodes within 2 years prior to the HSCT:

- 1 □ known
- 2 □ not known

52. Total number of episodes within the recipient’s lifetime:

- 1 □ known
- 2 □ not known

53. Did the recipient require exchange transfusion?

- 1 □ yes
- 2 □ no
- 3 □ unknown

Specify any treatment(s) the recipient required for acute chest syndrome:

54. 1 □ yes 2 □ no 3 □ unknown antibiotics

55. 1 □ yes 2 □ no 3 □ unknown intubation / mechanical ventilation

56. 1 □ yes 2 □ no 3 □ unknown oxygen

57. 1 □ yes 2 □ no 3 □ unknown transfusion of red blood cells

58. 1 □ yes 2 □ no 3 □ unknown other treatment

59. Specify: ________________________________
60. Osteonecrosis

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

Specify joint(s) affected:

61. Ankle
62. Hip
63. Knee
64. Shoulder
65. Spine
66. Other

67. Specify:

68. Priapism

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

69. Number of episodes experienced in the last 2 years:

- Known
- Not known

70. Was surgery performed to correct blood flow?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

71. Seizures

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

72. Sickle nephropathy

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

73. Stroke

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

74. Specify the total number of strokes:

- 1
- ≥2
- Unknown

75. Vaso-occlusive pain requiring hospitalization within 2 years prior to the HSCT

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

76. Specify the frequency of hospitalization:

- < 3 instances per year
- ≥3 instances per year
- Unknown

77. Did the recipient receive hydroxyurea at any time prior to the HSCT?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

78. Date hydroxyurea started:

- Month
- Day
- Year

79. Date hydroxyurea stopped:

- Month
- Day
- Year

80. Was hemoglobin electrophoresis performed while the recipient was receiving hydroxyurea?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

81. Date of electrophoresis:

- Month
- Day
- Year

If the recipient received chronic transfusions prior to HSCT, provide pre-transfusion electrophoresis data.

82. Date hydroxyurea started:

- Month
- Day
- Year

83. Date hydroxyurea stopped:

- Month
- Day
- Year

84. Was hemoglobin electrophoresis performed while the recipient was receiving hydroxyurea?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

85. Date of electrophoresis:

- Month
- Day
- Year

86. Date unknown
91. Did the recipient experience gonadal dysfunction at any time prior to the preparative regimen?
   1 □ yes
   2 □ no
   3 □ unknown

92. Was a brain MRI / MRA performed just prior to the preparative regimen?
   1 □ yes
   2 □ no
   3 □ unknown

93. Specify the MRI / MRA results:
   1 □ normal
   2 □ abnormal
   3 □ unknown

94. Is a copy of the MRI / MRA report attached to this form?
   1 □ yes
   2 □ no

95. Was an EKG performed prior to the preparative regimen?
   1 □ yes
   2 □ no
   3 □ unknown

96. Specify the EKG results:
   1 □ normal
   2 □ abnormal
   3 □ unknown

97. Is a copy of the EKG report attached to this form?
   1 □ yes
   2 □ no
98. Was an echocardiogram performed prior to the preparative regimen?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
</table>

99. Specify the echocardiogram results:

<table>
<thead>
<tr>
<th>Normal</th>
<th>Abnormal</th>
<th>Unknown</th>
</tr>
</thead>
</table>

100. Is a copy of the echocardiogram report attached to this form?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
</table>

101. Was the recipient's serum ferritin level tested at any time prior to the preparative regimen?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
</table>

102. Specify the serum ferritin results:

<table>
<thead>
<tr>
<th>&lt; 1,000 ng/mL or µg/L</th>
<th>≥ 1,001 ng/mL or µg/L</th>
<th>Unknown</th>
</tr>
</thead>
</table>

103. Was hemoglobin electrophoresis performed just prior to the preparative regimen (not including any electrophoresis reported in question 80)?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
</tr>
</thead>
</table>

If the recipient received chronic transfusions prior to HSCT, provide pre-transfusion electrophoresis data.

104. Date:

<table>
<thead>
<tr>
<th>Month</th>
<th>Day</th>
<th>Year</th>
<th>Unknown</th>
</tr>
</thead>
</table>

Specify the level of each hemoglobin type:

105. Hb A1: %

<table>
<thead>
<tr>
<th>%</th>
<th>Not tested</th>
</tr>
</thead>
</table>

106. Hb A2: %

<table>
<thead>
<tr>
<th>%</th>
<th>Not tested</th>
</tr>
</thead>
</table>

107. Hb C: %

<table>
<thead>
<tr>
<th>%</th>
<th>Not tested</th>
</tr>
</thead>
</table>

108. Hb F: %

<table>
<thead>
<tr>
<th>%</th>
<th>Not tested</th>
</tr>
</thead>
</table>

109. Hb S: %

<table>
<thead>
<tr>
<th>%</th>
<th>Not tested</th>
</tr>
</thead>
</table>

110. Other hemoglobin type

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
</table>

111. Specify type: ______________________

112. Level: %

113. Is a copy of the hemoglobin electrophoresis report attached to this form?

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
</table>
114. What was the primary reason for the HSCT?

1. [ ] acute chest syndrome
2. [ ] excessive transfusion requirements / iron overload
3. [ ] recurrent priapism
4. [ ] recurrent vaso-occlusive pain
5. [ ] stroke
6. [ ] other reason
7. [ ] unknown

115. Specify primary reason for HSCT: ____________________________________________

116. Signed: ________________________________________________________________

[ ] Person completing form

Please print name: ____________________________________________________________

Phone: (________________) ________________________________

Fax: (___________) __________________________________________________________

E-mail address: ______________________________________________________________