CIBMTR®
CENTER FOR INTERNATIONAL BLOOD & MARROW TRANSPLANT RESEARCH

Sickle Cell Anemia Pre-HSCT Data

CIBMTR Center Number: | | | | | |

CIBMTR Recipient ID: | | | | | | | | | | |

Today's Date: | | (Month) | | | (Day) | 2 0 | | (Year)

Date of HSCT for which this form is being completed: 🕮 | | (Month) | | | (Day) | 2 0 | | (Year)

HSCT type: ☐ autologous ☐ allogeneic, unrelated ☐ allogeneic, related ☐ syngeneic (identical twin)

Product type: ☐ marrow ☐ PBSC ☐ cord blood ☐ other product, specify: _____

Registry Use Only

Sequence Number:

Date Received:

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? | | (Month) | | | (Day) | | | | | (Year)

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 beta^0 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: | | (Month) | | | (Day) | | | | | (Year) ☐ 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

Mail this form to your designated campus (Milwaukee or Minneapolis). Retain the original at the transplant center.

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

10. 1 ☐ yes 2 ☐ no 3 ☐ unknown Duffy –Fy^a
11. 1 ☐ yes 2 ☐ no 3 ☐ unknown Kell –K
12. 1 ☐ yes 2 ☐ no 3 ☐ unknown Kell –k
13. 1 ☐ yes 2 ☐ no 3 ☐ unknown Kidd –Jk^a
14. 1 ☐ yes 2 ☐ no 3 ☐ unknown Kidd –Jk^b
15. 1 ☐ yes 2 ☐ no 3 ☐ unknown Lewis –Le^a
16. 1 ☐ yes 2 ☐ no 3 ☐ unknown Lewis –Le^b
17. 1 ☐ yes 2 ☐ no 3 ☐ unknown MNSs –M
18. 1 ☐ yes 2 ☐ no 3 ☐ unknown MNSs –N
19. 1 ☐ yes 2 ☐ no 3 ☐ unknown MNSs –S
20. 1 ☐ yes 2 ☐ no 3 ☐ unknown MNSs –s
21. 1 ☐ yes 2 ☐ no 3 ☐ unknown Rh –C
22. 1 ☐ yes 2 ☐ no 3 ☐ unknown Rh –D
23. 1 ☐ yes 2 ☐ no 3 ☐ unknown Rh –E
24. 1 ☐ yes 2 ☐ no 3 ☐ unknown Rh –e
25. 1 ☐ yes 2 ☐ no 3 ☐ unknown Rh –hr^a
26. 1 ☐ yes 2 ☐ no 3 ☐ unknown other → 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 → 33. Specify other route: _____
        - 6 ☐ unknown

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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

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Sickle Chronic Lung Disease Staging Criteria (Clinical)

Markers	Stage 1	Stage 2	Stage 3	Stage 4
Chest Pain	Recurrent substernal pain and chronic cough	Increased pain over Stage 1	Severe midline crushing chest pain	Severe and prolonged pain with dyspnea at rest
Blood Gasses	Normal oxygen saturation	Normal oxygen saturation	Hypoxia with partial pressure oxygen (70 mm Hg) during stable periods	Partial pressure oxygen (60 mm Hg) during stable periods
X-Ray	Decreased distal pulmonary vascularity, hyperexpansion, evidence suggestive of increased interstitial markings	Diffuse, fine interstitial fibrosis	Pulmonary fibrosis	Severe pulmonary fibrosis
Pulmonary Function Tests *	Decreased FVC, TLC, FEV_1 and FEV_1 / FVC ratio (mild, 80% of predicted normal, or 1 standard deviation below normal)	Decreased FVC, FEV_1, TLC, DCD and FEV_1 / FVC ratio (moderate, 60% of predicted, or 2 standard deviations below normal)	Decreased FVC, REV_1, TLC, DCO and FEV_1 / FVC ratio (severe, 40% of predicted, or 3 standard deviations below normal)	Patient frequently unable to complete testing due to degree of hypoxia
ECG and ECHO	Left ventricular preponderance persists	Balanced ventricular hypertrophy	Right ventricular hypertrophy and right atrial enlargement. Progressive increase in heart size	Severe right ventricular and right atrial hypertrophy. Ischemic T waves in V1 and V2, and P pulmonale
Pulmonary Artery Pressure	Normal	Normal	Borderline elevation or normal	Markedly elevated with pulmonary hypertension

***** These measurements are based on common methods for comparison of reference values.
Abbreviations: FVC = forced vial capacity, TLC = total lung capacity, REV_1 = 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: _____

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60. Osteonecrosis
- 1 ☐ yes → Specify joint(s) affected:
- 2 ☐ no
- 3 ☐ unknown

61. 1 ☐ yes 2 ☐ no 3 ☐ unknown ankle
62. 1 ☐ yes 2 ☐ no 3 ☐ unknown hip
63. 1 ☐ yes 2 ☐ no 3 ☐ unknown knee
64. 1 ☐ yes 2 ☐ no 3 ☐ unknown shoulder
65. 1 ☐ yes 2 ☐ no 3 ☐ unknown spine
66. 1 ☐ yes 2 ☐ no 3 ☐ unknown other → 67. Specify: _____

68. Priapism
- 1 ☐ yes → 69. Number of episodes experienced in the last 2 years:
  - 1 ☐ known → | | | | |
  - 2 ☐ not known
- 2 ☐ no
- 3 ☐ unknown

70. Was surgery performed to correct blood flow?
- 1 ☐ yes
- 2 ☐ no
- 3 ☐ unknown

71. Seizures
- 1 ☐ yes
- 2 ☐ no
- 3 ☐ unknown

72. Sickle nephropathy
- 1 ☐ yes
- 2 ☐ no
- 3 ☐ unknown

73. Stroke
- 1 ☐ yes → 74. Specify the total number of strokes:
  - 1 ☐ 1
  - 2 ☐ ≥ 2
  - 3 ☐ unknown
- 2 ☐ no
- 3 ☐ unknown

75. Vaso-occlusive pain requiring hospitalization within 2 years prior to the HSCT
- 1 ☐ yes → 76. Specify the frequency of hospitalization:
  - 1 ☐ < 3 instances per year
  - 2 ☐ ≥ 3 instances per year
  - 3 ☐ unknown
- 2 ☐ no
- 3 ☐ unknown

77. Did the recipient receive hydroxyurea at any time prior to the HSCT?
- 1 ☐ yes →
  - 78. Date hydroxyurea started: | | Month | | | Day | | | | Year | ☐ date unknown
  - 79. Date hydroxyurea stopped: | | Month | | | Day | | | | Year | ☐ date unknown
  - 80. Was hemoglobin electrophoresis performed while the recipient was receiving hydroxyurea?
    - 1 ☐ yes →
      - **If the recipient received chronic transfusions prior to HSCT, provide pre-transfusion electrophoresis data.**
      - 81. Date of electrophoresis: | | Month | | | Day | | | | Year | ☐ date unknown
    - 2 ☐ no
    - 3 ☐ unknown
- 2 ☐ no
- 3 ☐ unknown

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Specify the level of each hemoglobin type:

82. Hb A1: | | % ☐ not tested while receiving hydroxyurea

83. Hb A2: | | % ☐ not tested while receiving hydroxyurea8

84. Hb C: | | % ☐ not tested while receiving hydroxyurea

85. Hb F: | | % ☐ not tested while receiving hydroxyurea

86. Hb S: | | % ☐ not tested while receiving hydroxyurea

87. Other hemoglobin
- 1 ☐ yes → 88. Specify type: _____ 89. Level: | | %
- 2 ☐ no

90. Is a copy of the electrophoresis report attached?
- 1 ☐ yes
- 2 ☐ no

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 a 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

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98. Was an echocardiogram performed prior to the preparative regimen?

1 ☐ yes →
2 ☐ no
3 ☐ unknown

99. Specify the echocardiogram results:
1 ☐ normal
2 ☐ abnormal
3 ☐ unknown

100. Is a copy of the echocardiogram report attached to this form?
1 ☐ yes
2 ☐ no

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

1 ☐ yes →
2 ☐ no
3 ☐ unknown

102. Specify the serum ferritin results:
1 ☐ < 1,000 ng/mL or µg/L
2 ☐ ≥ 1,001 ng/mL or µg/L
3 ☐ unknown

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

1 ☐ yes →
2 ☐ no
3 ☐ unknown

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

104. Date : | | Month | | Day | | | | Year ☐ date unknown

Specify the level of each hemoglobin type:

105. Hb A1: | | % ☐ not tested

106. Hb A2: | | % ☐ not tested

107. Hb C: | | % ☐ not tested

108. Hb F: | | % ☐ not tested

109. Hb S: | | % ☐ not tested

110. Other hemoglobin type

1 ☐ yes →
2 ☐ no

111. Specify type: _____

112. Level: | | %

113. Is a copy of the hemoglobin electrophoresis report attached to this form?
1 ☐ yes
2 ☐ no

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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 → 115. Specify primary reason for HSCT: _____
7 ☐ unknown

116. Signed: _____
Person completing form

Please print name: _____

Phone: (_____) _____

Fax: (_____) _____

E-mail address: _____