**Form 2030 R2.0: Sickle Cell Anemia Pre-HSCT Data**

**Key Fields**

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

<table>
<thead>
<tr>
<th>CIBMTR Center Number:</th>
<th>CIBMTR Recipient ID:</th>
</tr>
</thead>
<tbody>
<tr>
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</table>

<table>
<thead>
<tr>
<th>Month</th>
<th>Day</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>0</td>
<td>20</td>
</tr>
</tbody>
</table>

**HSCT type (check all that apply):**

- Autologous
- Allogeneic, unrelated
- Allogeneic, related
- Syngeneic (identical twin)

**Product type (check all that apply):**

- Marrow
- PBSC
- Cord blood
- Other product

Specify: ____________________________

*If this is a report of a second or subsequent transplant, check here and continue with question 95.*

**Sickle Cell Anemia Pre-HSCT Data Questions: 1 - 118**

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

<table>
<thead>
<tr>
<th>Date of diagnosis:</th>
</tr>
</thead>
<tbody>
<tr>
<td>__ __ __ __ - __ __- __ __</td>
</tr>
</tbody>
</table>

2. **Was the recipient diagnosed with sickle cell disease at birth (i.e., newborn screening)?**

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

3. **What is the recipient's sickle cell disease genotype?**

| Hb SS |
| Hb S beta^thalassemia |
| Hb SC |
| Hb S beta^+ thalassemia |
| other genotype |

4. **Specify other genotype:**

Specify: ____________________________

5. **Did the recipient receive red blood cell transfusions at any time prior to the preparative regimen?**

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

6. **Date of first transfusion:**

| Date of first transfusion: |
| __ __ __ __ - __ __- __ __ |

<table>
<thead>
<tr>
<th>Date of first transfusion unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date of first transfusion unknown</td>
</tr>
</tbody>
</table>
### Specify the total number of transfusions received prior to the preparative regimen:

- < 5
- 5-10
- >10

### Did the transfusion(s) induce red cell alloimmunization?

- Yes
- No
- Unknown

### Specify the number of alloantibodies detected:

- 1
- >2
- Unknown

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

- 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

Mail, fax or email this form to Minneapolis. Fax: 612-627-5895. Email: scanform@nmdp.org. Retain the original form at the transplant center.
<table>
<thead>
<tr>
<th>Question</th>
<th>Yes</th>
<th>No</th>
<th>Unknown</th>
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<tbody>
<tr>
<td>23 Rh-E</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>24 Rh-e</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>25 Rh-h*</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>26 Other</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>27 Specify:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>28 Are red cell autoantibodies present?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>29 Specify the number of autoantibodies detected:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30 Was iron chelation therapy performed at any time prior to the regimen?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>31 Date chelation therapy started:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>32 Specify the predominant route of administration:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>33 Specify other route:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>34 Was a liver biopsy performed at any time prior to the preparative regimen?</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>35 Date of most recent liver biopsy:</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>36 Was hepatitis present?</td>
<td></td>
<td></td>
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<tr>
<td>37 Specify the severity of hepatitis:</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>38 Was siderosis present?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>39 Specify the severity of siderosis:</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>40 Was fibrosis present?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>41 Specify the severity of fibrosis:</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
42 Were serial liver biopsies performed?

- Yes
- No

43 Did the liver biopsies show progressive disease?

- Yes
- No
- Unknown

44 What was the hepatic iron concentration (HIC)?

- Known
- Unknown

45 Specify HIC: __________________ mg/g

46 Is a copy of the biopsy report attached?

- Yes
- No

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

- Yes
- No
- Unknown

48 Specify PFT results:

- Normal
- Stage 1 disease
- Stage 2 disease
- Stage 3 disease
- Stage 4 disease
- Unknown

49 Is a copy of the PFT report attached?

- Yes
- No

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

50 Acute chest syndrome

- Yes
- No
- Unknown

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

- Known
- Not known

52 Number of episodes: __________________

53 Total number of episodes within the recipient’s lifetime:

- Known
- Not known

54 Number of episodes: __________________

55 Did the recipient require exchange transfusion?

- Yes
- No
- Unknown

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

56 Antibiotics

- Yes
- No
- Unknown

57 Intubation / mechanical ventilation

- Yes
- No
- Unknown

58 Oxygen

- Yes
- No
- Unknown
<p>| | | | | | |</p>
<table>
<thead>
<tr>
<th></th>
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</thead>
<tbody>
<tr>
<td>59</td>
<td>transfusion of red blood cells</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>60</td>
<td>other treatment</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>61</td>
<td>Specify:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>62</td>
<td>Osteonecrosis</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>63</td>
<td>ankle</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>64</td>
<td>hip</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>65</td>
<td>knee</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>66</td>
<td>shoulder</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
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<tr>
<td>67</td>
<td>spine</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
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<tr>
<td>68</td>
<td>Other</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>69</td>
<td>Specify:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>70</td>
<td>Priapism</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>71</td>
<td>Number of episodes experienced in the last 2 years:</td>
<td>Known</td>
<td>Not known</td>
<td></td>
<td></td>
</tr>
<tr>
<td>72</td>
<td>Number of episodes:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>73</td>
<td>Was surgery performed to correct blood flow?</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>74</td>
<td>Seizures</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>75</td>
<td>Sickle nephropathy</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
<tr>
<td>76</td>
<td>Stroke</td>
<td>yes</td>
<td>no</td>
<td>Unknown</td>
<td></td>
</tr>
</tbody>
</table>
77 Specify the total number of strokes:
- 1
- >=2
- Unknown

78 Vaso-occlusive pain requiring hospitalization within 2 years prior to the HSCT
- Yes
- No
- Unknown

79 Specify the frequency of hospitalization:
- <3 instances per year
- >=3 instances per year
- Unknown

80 Did the recipient receive hydroxyurea at any time prior to the HSCT?
- Yes
- No
- Unknown

81 Date hydroxyurea started: __ __ __ __ - __ __ __ __
- Date hydroxyurea started unknown

82 Date hydroxyurea stopped: __ __ __ __ - __ __ __ __
- Date hydroxyurea stopped unknown

83 Was hemoglobin electrophoresis performed while the recipient was receiving hydroxyurea?
- Yes
- No
- Unknown

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

84 Date of electrophoresis: __ __ __ __ - __ __ __ __
- Date of electrophoresis unknown

Specify the level of each hemoglobin type:

85 Hb A1: ____________ %
- Hb A1 not tested while receiving hydroxyurea

86 Hb A2: ____________ %
- Hb A2 not tested while receiving hydroxyurea

87 Hb C: ____________ %
- Hb C not tested while receiving hydroxyurea

88 Hb F: ____________ %
- Hb F not tested while receiving hydroxyurea

89 Hb S: ____________ %
- Hb S not tested while receiving hydroxyurea

90 Other hemoglobin
- Yes
- No

91 Specify type: ________________

92 Level: ____________ %

93 Is a copy of the electrophoresis report attached?
- Yes
- No

94 Did the recipient experience gonadal dysfunction at any time prior to the preparative regimen?
- Yes
- No
- Unknown

95 Was a brain MRI / MRA performed just prior to the preparative regimen?
- Yes
- No
- Unknown

96 Specify the MRI / MRA results:
- Normal
- Abnormal
- Unknown
97 Is a copy of the MRI / MRA report attached to this form?

   yes  
   no

98 Was a EKG performed prior to the preparative regimen?

   yes  
   no  
   Unknown

99 Specify the EKG results:

   Normal  
   Abnormal  
   Unknown

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

   yes  
   no

101 Was an echocardiogram performed prior to the preparative regimen?

   yes  
   no  
   Unknown

102 Specify the echocardiogram results:

   Normal  
   Abnormal  
   Unknown

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

   yes  
   no

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

   yes  
   no  
   Unknown

105 Specify the serum ferritin results:

   <1,000 ng/mL or µg/L  
   >=1,001 ng/mL or µg/L  
   Unknown

106 Was hemoglobin electrophoresis performed just prior to the preparative regimen (not including any electrophoresis reported in question 83)?

   yes  
   no  
   Unknown

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

107 Date: __ __ __ __ - __ __ - __ __  

108 Specify the level of each hemoglobin type:

   Hb A1:  %  
   Hb A1 not tested

   Hb A2:  %  
   Hb A2 not tested

   Hb C:  %  
   Hb C not tested

   Hb F:  %  
   Hb F not tested

   Hb S:  %  
   Hb S not tested

110 Other hemoglobin type

   yes  
   no

114 Specify type: ______________________

115 Level: ______________________  %
116 Is a copy of the hemoglobin electrophoresis report attached to this form?
   yes  no

117 What was the primary reason for the HSCT?
   acute chest syndrome
   excessive transfusion requirements / iron overload
   recurrent priapism
   recurrent vaso-occlusive pain
   stroke
   other reason
   Unknown

118 Specify primary reason for HSCT: ________________________________

First Name: ___________________________ Last Name: ___________________________
Phone number: _______________________ Fax number: _______________________
E-mail address: ______________________