Has this patient’s data been previously reported to USIDNET?

- [ ] yes
- [x] no

Laboratory Studies Post-HSCT

Report the most recent findings since the date of the last report. For questions 1–3 and 6–7, also report CBC results in the Form 2100 – 100 Days Post-HSCT Data beginning at question 48, or in the Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 19.

1. Date of most recent hematologic testing: [ ] Month [ ] Day [ ] Year

Specify units:

1. [ ] x 10^9/L (x 10^3/mm^3)
2. [x] x 10^6/L

2. WBC:

Specify units:

3. [ ] %
4. [ ] %

5. Polymorphonuclear leukocytes (PMN):

Specify units:

6. Hemoglobin:

Specify units:

7. Platelets:

Specify units:

8. Mean platelet volume:

Specify units:

9. What was the platelet size at the date of the most recent follow-up?

- [ ] decreased
- [ ] normal
- [ ] unknown

Visit: [ ] 100 day [ ] 6 month [ ] 1 year [ ] 2 years [ ] > 2 years, specify: [ ]
Immunoglobulin Analysis
Specify the most recent quantitative immunoglobulins measured since the date of the last report. For questions 10–15, also report immunoglobulins in the Form 2100 – 100 Days Post-HSCT Data beginning at question 55, or in the Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 26. For questions 18–19, also report IVIG in the Form 2100 – 100 Days Post-HSCT Data beginning at question 61, or in the Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 32.

10. IgG: 11. o not tested
12. IgM: 13. o not tested
14. IgA: 15. o not tested
16. IgE: IU/mL 17. o not tested

18. Did the recipient receive supplemental intravenous immunoglobulins (IVIG) since the date of the last report?
   1 o yes
   2 o no
   3 o unknown

Lymphocyte Analysis
Specify the most recent lymphocyte assessment measured since the date of the last report. For questions 21 and 23–27, also report lymphocytes in the Form 2100 – 100 Days Post-HSCT Data beginning at question 71, or in the Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 42.

20. Were lymphocyte analyses performed?
   1 o yes
   2 o no

21. Date of most recent testing performed:
   Month Day Year
22. Absolute lymphocyte count:
   cells / µL (cells / mm³)
23. CD3 (T cells):
   % of total lymphocytes: Value:
   Specify units:
   1 o x 10⁹/L (x 10³/mm³) o not tested
   2 o x 10⁶/L o not tested
24. CD4 (T helper cells):
   % of total lymphocytes: Value:
   Specify units:
   1 o x 10⁹/L (x 10³/mm³) o not tested
   2 o x 10⁶/L o not tested
25. CD8 (cytotoxic T cells):
   % of total lymphocytes: Value:
   Specify units:
   1 o x 10⁹/L (x 10³/mm³) o not tested
   2 o x 10⁶/L o not tested
26. CD20 (B lymphocyte cells):
   % of total lymphocytes: Value:
   Specify units:
   1 o x 10⁹/L (x 10³/mm³) o not tested
   2 o x 10⁶/L o not tested
27. CD56 (natural killer (NK) cells):
   % of total lymphocytes: Value:
   Specify units:
   1 o x 10⁹/L (x 10³/mm³) o not tested
   2 o x 10⁶/L o not tested
Antibody Response
Specify the most recent antibody responses measured since the date of the last report.

30. Date antibody responses were assessed: __ __ __ 20 __ __ __

<table>
<thead>
<tr>
<th>Antibody Response</th>
<th>Absent</th>
<th>Low</th>
<th>Normal</th>
<th>Not tested</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>3</td>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

31. Bacteriophage phi X-174 or other neoantigen
32. Diptheria
33. Isohemagglutinin anti-A
34. Isohemagglutinin anti-B
35. Protein conjugated HIB or pneumococcal vaccine
36. Tetanus

37. Unconjugated pneumococcal polysaccharide: ___ / ___
   Number of serotypes producing a protective level / Total serotypes tested from vaccine

38. Conjugated pneumococcal polysaccharide: ___ / ___
   Number of serotypes producing a protective level / Total serotypes tested from vaccine

Lymphocyte Function
Specify the most recent lymphocyte function measured since the date of the last report.

39. Date lymphocyte function was assessed: __ __ __ 20 __ __ __

<table>
<thead>
<tr>
<th>Lymphocyte Function</th>
<th>Absent (&lt;10% of control)</th>
<th>Low (10-30% of control)</th>
<th>Normal (&gt;30% of control)</th>
<th>Not tested</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>3</td>
<td>4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

40. Anti-CD3
41. Candida antigen
42. Concanavalin A (ConA)
43. Phytohemagglutinin (PHA)
44. Pokeweed mitogen (PWM)
45. Tetanus antigen

46. What is the current natural killer cell function? (Refers to specific cytolysis of NK-sensitive target cells, e.g. K562.)
   1  absent (≤ 10% normal response)
   2  decreased (11–50% normal response)
   3  normal
   4  unknown
47. Did a new malignancy, lymphoproliferative or myeloproliferative disorder appear that is different from the disease for which the HSCT was performed?

1  yes
2  no

48. Specify second malignancy:
1  EBV-associated B-cell lymphoproliferative disorder
2  other second malignancy
3  unknown

49. Specify other second malignancy:

50. Specify the date of diagnosis:

Also report malignancy in the Form 2100 – 100 Days Post-HSCT Data beginning at question 519, Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 459, or Form 2300 — Yearly Follow-Up for Greater Than Two Years Post-HSCT Data beginning at question 131. Copy questions 46–49 to report more than one secondary malignancy; check here if additional pages are attached.

Clinical Status of Recipient Post-HSCT

51. Did the recipient experience any types of bleeding (since the date of the last report)?

1  yes
2  no

Specify types of bleeding:

Bleeding episode(s) present?

If present, is the feature prominent?

52. 1  yes 2  no Epistaxis

53. 1  yes 2  no

54. 1  yes 2  no Upper GI hemorrhage

55. 1  yes 2  no

56. 1  yes 2  no Lower GI hemorrhage / rectal bleeding

57. 1  yes 2  no

58. 1  yes 2  no Hemarthrosis

59. 1  yes 2  no

60. 1  yes 2  no Hematuria

61. 1  yes 2  no

62. 1  yes 2  no Intracranial hemorrhage

63. 1  yes 2  no

64. 1  yes 2  no Oral

65. 1  yes 2  no

66. 1  yes 2  no Subcutaneous bleeding

67. 1  yes 2  no

68. 1  yes 2  no Subdural hematoma

69. 1  yes 2  no

70. 1  yes 2  no Other bleeding

71. 1  yes 2  no

72. Specify other bleeding:

73. Did the recipient experience any autoimmune / inflammatory disorders (since the date of the last report)?

1  yes
2  no

Specify autoimmune / inflammatory disorders:

Feature present?

If present, is the feature prominent?

74. 1  yes 2  no Arthralgia

75. 1  yes 2  no

76. 1  yes 2  no Arthritis, chronic

77. 1  yes 2  no

78. 1  yes 2  no Autoimmune hemolytic anemia

79. 1  yes 2  no

80. 1  yes 2  no Idiopathic thrombocytopenic purpura (ITP)

81. 1  yes 2  no

82. 1  yes 2  no Inflammatory bowel disease

83. 1  yes 2  no

84. 1  yes 2  no Juvenile rheumatoid arthritis

85. 1  yes 2  no

86. 1  yes 2  no Nephritis

87. 1  yes 2  no

88. 1  yes 2  no Neutropenia

89. 1  yes 2  no

90. 1  yes 2  no Sclerosing cholangitis

91. 1  yes 2  no

92. 1  yes 2  no Vasculitis, cerebral

93. 1  yes 2  no

94. 1  yes 2  no Vasculitis, coronary

95. 1  yes 2  no

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
Post-HSCT Treatment for Wiskott-Aldrich Syndrome

106. Was any treatment given for relapsed, persistent, or progressive disease (since the date of the last report)?

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>108. yes</td>
<td>10. 20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

107. Antithymocyte globulin (ATG, ATGAM, Thymoglobulin)

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>109. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

108. Corticosteroids, systemic

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>112. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

109. Corticosteroids, topical

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>115. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

110. Cyclophosphamide (CTX, Cytoxan, Neosar)

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>118. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

111. Cyclosporine (CsA, Neoral, Sandimmune)

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>121. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

112. In vivo monoclonal antibody

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>125. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Specify monoclonal antibody:

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>128. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

120. Daclizumab (anti-CD25, Zenapax)

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>131. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

121. Etanercept (Enbrel)

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>130. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

122. In vivo monoclonal antibody

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Therapy Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 yes</td>
<td>124. yes</td>
<td>20</td>
</tr>
<tr>
<td>2 no</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Also report immunosuppressive medications given to prevent or treat GVHD in the corresponding questions on the Form 2000 — Recipient Baseline Data, Form 2100 — 100 Days Post-HSCT Data, Form 2200 — Six Months to Two Years Post-HSCT Data, or Form 2300 — Yearly Follow-Up for Greater Than Two Years Post-HSCT Data.

Therapy paused for < 1 week should not be considered as “Therapy Stopped.”
<table>
<thead>
<tr>
<th>Therapy Given</th>
<th>Therapy Stopped</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>132. Infliximab (anti-TNF-α, Remicade)</td>
<td>133. yes</td>
<td>134. yes</td>
</tr>
<tr>
<td>143. Muromonab (anti-CD3, OKT3)</td>
<td>144. yes</td>
<td>145. yes</td>
</tr>
<tr>
<td>149. Rituximab (anti-CD20, Rituxan, MabThera)</td>
<td>150. yes</td>
<td>151. yes</td>
</tr>
<tr>
<td>152. Other monoclonal antibody</td>
<td>153. yes</td>
<td>154. yes</td>
</tr>
<tr>
<td>155. Sirolimus (Rapamune)</td>
<td>156. yes</td>
<td>157. yes</td>
</tr>
<tr>
<td>158. Tacrolimus (FK506, Prograf)</td>
<td>159. yes</td>
<td>160. yes</td>
</tr>
<tr>
<td>161. Thalidomide (Thalomid)</td>
<td>162. yes</td>
<td>163. yes</td>
</tr>
<tr>
<td>164. Other immunosuppressive drug</td>
<td>165. yes</td>
<td>166. yes</td>
</tr>
</tbody>
</table>

167. Did the recipient receive any other significant treatment(s) for WAS (since the date of the last report)?

168. Specify other treatment(s):
169. What is the current status of T-cell engraftment?

1. Predominantly or completely donor (≥ 80% donor chimerism)
2. Mixed chimerism (5–80% donor)
3. Only host T-cells detected (< 5% donor)
4. Unknown

170. Most recent date T-cell engraftment was assessed: [date unknown]

171. What is the current status of B-cell engraftment?

1. Predominantly or completely donor (≥ 80% donor chimerism)
2. Mixed chimerism (5–80% donor)
3. Only host B-cells detected (< 5% donor)
4. Unknown

172. Most recent date B-cell engraftment was assessed: [date unknown]

173. What is the current status of myeloid engraftment?

1. Predominantly or completely donor (≥ 80% donor chimerism)
2. Mixed chimerism (5–80% donor)
3. Only host myeloid cells detected (< 5% donor)
4. Unknown

174. Most recent date myeloid engraftment was assessed: [date unknown]

175. Signed: ____________________________

Person completing form

Please print name: ____________________________

Phone number: (__________) ____________________

Fax number: (__________) ______________________

E-mail address: ____________________________________

This section refers to quantitative analyses utilizing discriminating DNA markers. Peripheral blood cells must undergo separation or sorting into T, B, or lymphoid vs. myeloid populations to perform this determination. If RFLP analyses indicate only donor type hematopoiesis, mark T-cell, B-cell, and myeloid as “predominantly or completely donor.”

Also report chimerism in the Form 2100 — 100 Days Post-HSCT Data beginning at question 77 or Form 2200 — Six Months to Two Years Post-HSCT Data beginning at question 48.