CIBMTR®
CENTER FOR INTERNATIONAL BLOOD & MARROW TRANSPLANT RESEARCH

Wiskott-Aldrich Syndrome Post-HSCT Data

Registry Use Only

Sequence Number:

Date Received:

CIBMTR Center Number:

CIBMTR Recipient ID:

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

1 ☐ yes → USIDNET ID:
2 ☐ no

Today's Date: Month Day Year (20__)

Date of HSCT for which this form is being completed: 🕮 Month Day Year

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

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

Visit: ☐ 100 day ☐ 6 month ☐ 1 year ☐ 2 years ☐ > 2 years, specify:

To be completed in conjunction with a 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. Information reported here should reflect the date of last contact as reported in the post-HSCT data collection form, or immediately prior to death.

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 (20__)

Specify units:

2. WBC: ____.__ 1 ☐ x 10^9/L (x 10^3/mm^3) 2 ☐ x 10^6/L ☐ not tested

3. Lymphocytes: __ % ☐ not tested

4. Eosinophils: __ % ☐ not tested

5. Polymorphonuclear leukocytes (PMN): __ % ☐ not tested

6. Hemoglobin: ____.__ 1 ☐ g/dL 2 ☐ g/L 3 ☐ mmol/L ☐ not tested ☐ transfused RBC < 30 days from date of most current testing

7. Platelets: _____ 1 ☐ x 10^9/L (x 10^3/mm^3) 2 ☐ x 10^6/L ☐ not tested ☐ transfused platelets < 7 days from date of most current testing

8. Mean platelet volume: ___.__ fl ☐ not tested

9. What was the platelet size at the date of the most recent follow-up?
   1 ☐ decreased
   2 ☐ normal
   3 ☐ unknown

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

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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.

	Value:	Specify units:	Date tested: Month Day Year	
10. IgG:	☐☐☐☐☐ • ☐☐	1 ☐ mg/dL 2 ☐ g/dL 3 ☐ g/L	11. ☐☐ ☐☐ ☐☐☐☐	☐ not tested
12. IgM:	☐☐☐☐☐ • ☐☐	1 ☐ mg/dL 2 ☐ g/dL 3 ☐ g/L	13. ☐☐ ☐☐ ☐☐☐☐	☐ not tested
14. IgA:	☐☐☐☐☐ • ☐☐	1 ☐ mg/dL 2 ☐ g/dL 3 ☐ g/L	15. ☐☐ ☐☐ ☐☐☐☐	☐ not tested
16. IgE:	☐☐☐ IU/mL		17. ☐☐ ☐☐ ☐☐☐☐	☐ not tested

18. Did the recipient receive supplemental intravenous immunoglobulins (IVIG) since the date of the last report?
1 ☐ yes → 19. Was therapy ongoing within one month of immunoglobulin testing? 1 ☐ yes 2 ☐ no
2 ☐ no
3 ☐ 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 ☐ yes →
2 ☐ no

21. Date of most recent testing performed: ☐☐ ☐☐ ☐☐☐☐ (Month Day Year)

22. Absolute lymphocyte count: → ☐☐☐☐☐☐☐ cells / µL (cells / mm^3)

	% of total lymphocytes:		Value:	Specify units:	
23. CD3 (T cells): →	☐☐	– or –	☐☐☐☐☐☐☐	1 ☐ x 10^9/L ($x 10^3/mm^3$) 2 ☐ x 10^6/L	☐ not tested
24. CD4 (T helper cells): →	☐☐	– or –	☐☐☐☐☐☐☐	1 ☐ x 10^9/L ($x 10^3/mm^3$) 2 ☐ x 10^6/L	☐ not tested
25. CD8 (cytotoxic T cells): →	☐☐	– or –	☐☐☐☐☐☐☐	1 ☐ x 10^9/L ($x 10^3/mm^3$) 2 ☐ x 10^6/L	☐ not tested
26. CD20 (B lymphocyte cells): →	☐☐	– or –	☐☐☐☐☐☐☐	1 ☐ x 10^9/L ($x 10^3/mm^3$) 2 ☐ x 10^6/L	☐ not tested
27. CD56 (natural killer (NK) cells): →	☐☐	– or –	☐☐☐☐☐☐☐	1 ☐ x 10^9/L ($x 10^3/mm^3$) 2 ☐ x 10^6/L	☐ not tested

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	% of total lymphocytes:		Value:		Specify units:	
28. CD4+ / CD45RA+ (naive T cells): →		– or –			1 ☐ x 10^9/L (x 10^3/mm^3) 2 ☐ x 10^6/L	☐ not tested
29. CD4+ / CD45RO+ (memory T cells): →		– or –			1 ☐ x 10^9/L (x 10^3/mm^3) 2 ☐ x 10^6/L	☐ not tested

Antibody Response

Specify the most recent antibody responses measured since the date of the last report.

30. Date antibody responses were assessed: | | Month | | Day | 2 0 | Year

Absent	Low	Normal	Not tested	
1 ☐	2 ☐	3 ☐	4 ☐	31. Bacteriophage phi X-174 or other neoantigen
1 ☐	2 ☐	3 ☐	4 ☐	32. Diptheria
1 ☐	2 ☐	3 ☐	4 ☐	33. Isohemagglutinin anti-A
1 ☐	2 ☐	3 ☐	4 ☐	34. Isohemagglutinin anti-B
1 ☐	2 ☐	3 ☐	4 ☐	35. Protein conjugated HIB or pneumococcal vaccine
1 ☐	2 ☐	3 ☐	4 ☐	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: | | Month | | Day | 2 0 | Year

Absent (< 10% of control)	Low (10-30% of control)	Normal (> 30% of control)	Not tested	
1 ☐	2 ☐	3 ☐	4 ☐	40. Anti-CD3
1 ☐	2 ☐	3 ☐	4 ☐	41. Candida antigen
1 ☐	2 ☐	3 ☐	4 ☐	42. Concavalin A (ConA)
1 ☐	2 ☐	3 ☐	4 ☐	43. Phytohemagglutinin (PHA)
1 ☐	2 ☐	3 ☐	4 ☐	44. Pokeweed mitogen (PWM)
1 ☐	2 ☐	3 ☐	4 ☐	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

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47. Did a new malignancy, lymphoproliferative or myeloproliferative disorder appear that is different from the disease for which the HSCT was performed?

1 ☐ yes → 48. Specify second malignancy:
2 ☐ no

1 ☐ EBV-associated B-cell lymphoproliferative disorder
2 ☐ other second malignancy → 49. Specify other second malignancy: _____
3 ☐ unknown

50. Specify the date of diagnosis: | | Month | | | Day | 2 0 | | Year

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 → Specify types of bleeding:
2 ☐ no

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 → Specify autoimmune / inflammatory disorders:
2 ☐ no

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

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96. 1 ☐ yes 2 ☐ no Vasculitis, renal → 97. 1 ☐ yes 2 ☐ no
98. 1 ☐ yes 2 ☐ no Vasculitis, skin → 99. 1 ☐ yes 2 ☐ no
100. 1 ☐ yes 2 ☐ no Vasculitis, other → 101. 1 ☐ yes 2 ☐ no
102. Specify other vasculitis: _____
103. 1 ☐ yes 2 ☐ no Other disorder → 104. 1 ☐ yes 2 ☐ no
105. Specify other disorder: _____

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)?

1 ☐ yes → **Complete the table below.**

2 ☐ no → **Continue with question 169.**

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."

	Therapy Given?	Therapy Stopped?	Date Stopped (Month / Day / Year)	
107. Antithymocyte globulin (ATG, ATGAM, Thymoglobulin)	1 ☐ yes 2 ☐ no	108. 1 ☐ yes 2 ☐ no → 109.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
110. Corticosteroids, systemic	1 ☐ yes 2 ☐ no	111. 1 ☐ yes 2 ☐ no → 112.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
113. Corticosteroids, topical	1 ☐ yes 2 ☐ no	114. 1 ☐ yes 2 ☐ no → 115.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
116. Cyclophosphamide (CTX, Cytoxan, Neosar)	1 ☐ yes 2 ☐ no	117. 1 ☐ yes 2 ☐ no → 118.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
119. Cyclosporine (CsA, Neoral, Sandimmune)	1 ☐ yes 2 ☐ no	120. 1 ☐ yes 2 ☐ no → 121.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown

122. In vivo monoclonal antibody

1 ☐ yes → Specify monoclonal antibody:
2 ☐ no

	Therapy Given?	Therapy Stopped?	Date Stopped (Month / Day / Year)	
123. Alemtuzumab (Campath)	1 ☐ yes 2 ☐ no	124. 1 ☐ yes 2 ☐ no → 125.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
126. Daclizumab (anti-CD25, Zenapax)	1 ☐ yes 2 ☐ no	127. 1 ☐ yes 2 ☐ no → 128.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown
129. Etanercept (Enbrel)	1 ☐ yes 2 ☐ no	130. 1 ☐ yes 2 ☐ no → 131.	☐☐ / ☐☐ / 20☐☐	☐ date estimated ☐ date unknown

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Therapy Given?	Therapy Stopped?	Date Stopped (Month / Day / Year)	
132. Infliximab (anti-TNF-α, Remicade)	1 ☐ yes → 133. 1 ☐ yes → 134. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
135. Muromonab (anti-CD3, OKT3)	1 ☐ yes → 136. 1 ☐ yes → 137. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
138. Rituximab (anti-CD20, Rituxan, MabThera)	1 ☐ yes → 139. 1 ☐ yes → 140. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
141. Other monoclonal antibody	1 ☐ yes → 142. 1 ☐ yes → 143. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown

144. Specify other monoclonal antibody: _____

Therapy Given?	Therapy Stopped?	Date Stopped (Month / Day / Year)	
145. Lenalidomide (Revlimid)	1 ☐ yes → 146. 1 ☐ yes → 147. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
148. Mycophenolate mofetil (MMF, Cellcept)	1 ☐ yes → 149. 1 ☐ yes → 150. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
151. Photopheresis / extracorporeal phototherapy (ECP)	1 ☐ yes → 152. 1 ☐ yes → 153. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
154. Sirolimus (Rapamune)	1 ☐ yes → 155. 1 ☐ yes → 156. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
157. Tacrolimus (FK506, Prograf)	1 ☐ yes → 158. 1 ☐ yes → 159. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
160. Thalidomide (Thalomid)	1 ☐ yes → 161. 1 ☐ yes → 162. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown
163. Other immunosuppressive drug	1 ☐ yes → 164. 1 ☐ yes → 165. / 2 ☐ no / 2 ☐ no	☐☐ / ☐☐ / 2 0 ☐☐	☐ date estimated ☐ date unknown

166. Specify other immunosuppressive drug: _____

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

1 ☐ yes → 168. Specify other treatment(s): _____

2 ☐ no

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Status of Hematologic Engraftment

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.

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: | | Month | | | Day | | 2 | 0 | | | Year | ☐ 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: | | Month | | | Day | | 2 | 0 | | | Year | ☐ 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: | | Month | | | Day | | 2 | 0 | | | Year | ☐ date unknown

175. Signed: _____
Person completing form

Please print name: _____

Phone number: (_____) _____

Fax number: (_____) _____

E-mail address: _____

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