Form 2133 R3.0: Wiskott-Aldrich Syndrome Post-HSCT Data

Center: CRID:

Key Fields

Sequence Number: _____

Date Received: __ __ __ __ - __ __- __ __

CIBMTR Center Number: _____

CIBMTR Recipient ID: _____

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

yes no

USIDNET ID: _____

Today's Date: __ __ __ __ - __ __- __ __

Date of HSCT for which this form is being completed: __ __ __ __ - __ __- __ __

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 _____

Visit:

100 day 6 months 1 year 2 years > 2 years,

Specify: _____

Laboratory Studies Post-HSCT

Questions: 1 - 50

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: __ __ __ __ - __ __- __ __

2 WBC: _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

WBC not tested

3 Lymphocytes: _____ % Lymphocytes not tested

4 Eosinophils: _____ % Eosinophils not tested

5 Polymorphonuclear leukocytes (PMN): _____ % Polymorphonuclear leukocytes (PMN) not tested

6 Hemoglobin: _____ g/dL g/L mmol/L

Hemoglobin not tested

transfused RBC < 30 days from date of most current testing

7 Platelets: _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

Platelets not tested

transfused platelets < 7 days from date of most current testing

8 Mean platelet volume: _____ fl Mean platelet volume not tested

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

Decreased Normal Unknown

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: _____ mg/dL g/dL g/L

IgG not tested

11 Date tested: __ __ __ __ - __ __- __ __

12 IgM: _____ mg/dL g/dL g/L

IgM not tested

13 Date tested: __ __ __ __ - __ __- __ __

14 IgA: _____ mg/dL g/dL g/L

IgA not tested

15 Date tested: __ __ __ __ - __ __- __ __

16 IgE: _____ IU/mL

17 Date tested: __ __ __ __ - __ __- __ __ IgE not tested

18 Did the recipient receive supplemental intravenous immunoglobulins (IVIG)(since the date of the last report)?

yes no Unknown

19 Was therapy ongoing within one month of immunoglobulin testing?

yes no

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?

yes no

21 Date of most recent testing performed: __ __ __ __ - __ __- __ __

22 Absolute lymphocyte count: _____ cells / µL (cells / mm^3)

23 CD3 (T cells) % of total lymphocytes _____ %

- OR -

CD3 (T cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD3 (T cells) not tested

24 CD4 (T helper cells) % of total lymphocytes _____ %

- OR -

CD4 (T helper cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD4 (T helper cells) not tested

25 CD8 (cytotoxic T cells) % of total lymphocytes _____ %

- OR -

CD8 (cytotoxic T cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD8 (cytotoxic T cells) not tested

26 CD20 (B lymphocyte cells) % of total lymphocytes _____ %

- OR -

CD20 (B lymphocyte cells) value: _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD20 (B lymphocyte cells) not tested

27 CD56 (natural killer (NK) cells) % of total lymphocytes _____ %

- OR -

CD56 (natural killer (NK) cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD56 (natural killer (NK) cells) not tested

28 CD4+/CD45RA+ (naive T cells) % of total lymphocytes _____ %

- OR -

CD4+/CD45RA+ (naive T cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD4+/CD45RA+ (memory T cells) not tested

29 CD4+/CD45RO+ (memory T cells) % of total lymphocytes _____ %

- OR -

CD4+/CD45RO+ (memory T cells) value _____ x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD4+/CD45RO+ (memory T cells) not tested

Antibody Response

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

30 Date antibody responses were assessed: __ __ __ __ - __ __- __ __

31 Bacteriophage phi X-174 or other neoantigen

Absent Low Normal Not Tested

32 Diptheria

Absent Low Normal Not Tested

33 Isohemagglutinin anti-A

Absent Low Normal Not Tested

34 Isohemagglutinin anti-B

Absent Low Normal Not Tested

35 Protein conjugated HIB or pneumococcal vaccine

Absent Low Normal Not Tested

36 Tetanus

Absent Low Normal Not Tested

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: __ __ __ __ - __ __- __ __

40 Anti-CD3

Absent

Low (10-30% of control)

Normal

Not tested

41 Candida antigen

Absent

Low (10-30% of control)

Normal

Not tested

42 Concavalin A (ConA)

Absent

Low (10-30% of control)

Normal

Not tested

43 Phytohemagglutinin (PHA)

- Absent
- Low (10-30% of control)
- Normal
- Not tested

44 Pokeweed mitogen (PWM)

- Absent
- Low (10-30% of control)
- Normal
- Not tested

45 Tetanus antigen

- Absent
- Low (10-30% of control)
- Normal
- Not tested

46 What is the current natural killer cell function? (Refers to specific cytolysis of NK-sensitive target cells, e.g. K562.)

- absent (<= 10% normal response)
- decreased(11-50% normal response)
- Normal
- Unknown

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

- yes
- no

New Malignancy (1) **Questions: 48 - 50**

48 Specify second malignancy:

- EBV-associated B-cell lymphoproliferative disorder
- other second malignancy
- 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 47–50 to report more than one secondary malignancy;

- Check here if additional pages are attached.

Clinical Status of Recipient Post-HSCT

Questions: 51 - 105

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

- yes
- no

Specify types of bleeding:

52 Is epistaxis present?

yes no

53 Is epistaxis prominent?

yes no

54 Is upper GI hemorrhage present?

yes no

55 Is upper GI hemorrhage prominent?

yes no

56 Lower GI hemorrhage / rectal bleeding

yes no

57 Is lower GI hemorrhage/rectal bleeding prominent?

yes no

58 Is hemarthrosis present?

yes no

59 Is hemarthrosis prominent?

yes no

60 Is hematuria present?

yes no

61 Is hematuria prominent?

yes no

62 Is intracranial hemorrhage present?

yes no

63 Is intracranial hemorrhage prominent?

yes no

64 Is oral bleeding present?

yes no

65 Is oral bleeding prominent?

yes no

66 Is subcutaneous bleeding present?

yes no

67 Is subcutaneous bleeding prominent?

yes no

68 Is subdural hematoma present?

yes no

69 Is subdural hematoma prominent?

yes no

70 Is other bleeding present?

yes no

71 Is other bleeding prominent?

yes no

72 Specify other bleeding: _____

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

yes no

Specify autoimmune / inflammatory disorders:

74 Is arthralgia present?

yes no

75 Is arthralgia prominent?

yes no

76 Is chronic arthritis present?

yes no

77 Is chronic arthritis prominent?

yes no

78 Is autoimmune hemolytic anemia present?

yes no

79 Is autoimmune hemolytic anemia prominent?

yes no

80 Is idiopathic thrombocytopenic purpura (ITP) present?

yes no

81 Is idiopathic thrombocytopenic purpura (ITP) prominent?

yes no

82 Is inflammatory bowel disease present?

yes no

83 Is inflammatory bowel disease prominent?

yes no

84 Is juvenile rheumatoid arthritis present?

yes no

85 Is juvenile rheumatoid arthritis present?

yes no

86 Is nephritis present?

yes no

87 Is nephritis prominent?

yes no

88 Is neutropenia present?

yes no

89 Is neutropenia prominent?

yes no

90 Is sclerosing cholangitis present?

yes no

91 Is sclerosing cholangitis prominent?

yes no

92 Is cerebral vasculitis present?

yes no

93 Is cerebral vasculitis prominent?

yes no

94 Is coronary vasculitis present?

yes no

95 Is coronary vasculitis prominent?

yes no

96 Is renal vasculitis present?

yes no

97 Is renal vasculitis prominent?

yes no

98 Is skin vasculitis present?

yes no

99 Is skin vasculitis prominent?

yes no

100 Is other vasculitis present?

yes no

101 Is other vasculitis prominent?

yes no

102 Specify other vasculitis: _____

103 Is any other disorder present?

yes no

104 Is any other disorder prominent?

yes no

105 Specify other disorder: _____

Post-HSCT Treatment for Wiskott-Aldrich Syndrome

Questions: 106 - 168

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

yes no

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."

107 Antithymocyte globulin (ATG, ATGAM, Thymoglobulin)

yes no

108 Was therapeutic antithymocyte globulin (ATG, ATGAM, Thymoglobulin) stopped?

yes no

109 Date therapeutic antithymocyte globulin (ATG, ATGAM, Thymoglobulin) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

110 Were systemic corticosteroids given as therapy?

yes no

111 Were therapeutic systemic corticosteroids stopped?

yes no

112 Date therapeutic systemic corticosteroids stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

113 Were topical corticosteroids given as therapy?

yes no

114 Were therapeutic topical corticosteroids stopped?

yes no

115 Date therapeutic topical corticosteroids stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

116 Was cyclophosphamide (CTX, Cytoxan, Neosar) given as therapy?

yes no

117 Was therapeutic cyclophosphamide (CTX, Cytoxan, Neosar) stopped?

yes no

118 Date therapeutic cyclophosphamide (CTX, Cytoxan, Neosar) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

119 Was cyclosporine (CsA, Neoral, Sandimmune) given as therapy?

yes no

120 Was therapeutic cyclosporine (CsA, Neoral, Sandimmune) stopped?

yes no

121 Date therapeutic cyclosporine (CsA, Neoral, Sandimmune) stopped? __ __ __ __ - __ __- __ __ date estimated

Date unknown

122 Was in vivo monoclonal antibody given as therapy?

yes no

Specify monoclonal antibody:

123 Was alemtuzumab (Campath) given as therapy?

yes no

124 Was therapeutic alemtuzumab (Campath) stopped?

yes no

125 Date therapeutic alemtuzumab (Campath) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

126 Was daclizumab (anti-CD25, Zenapax) given as therapy?

yes no

127 Was therapeutic daclizumab (anti-CD25, Zenapax) stopped?

yes no

128 Date therapeutic daclizumab (anti-CD25, Zenapax) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

129 Was etanercept (Enbrel) given as therapy?

yes no

130 Was therapeutic etanercept (Enbrel) stopped?

yes no

131 Date therapeutic etanercept (Enbrel) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

132 Was infliximab (anti-TNF-α, Remicade) given as therapy?

yes no

133 Was therapeutic infliximab (anti-TNF-α, Remicade) stopped?

yes no

134 Date therapeutic infliximab (anti-TNF-α, Remicade) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

135 Was muromonab (anti-CD3, OKT3) given as therapy?

yes no

136 Was therapeutic muromonab (anti-CD3, OKT3) stopped?

yes no

137 Date therapeutic muromonab (anti-CD3, OKT3) stopped: __ __ __ __ - __ __- __ __ date estimated

Date unknown

138 Rituximab (anti-CD20, Rituxan, MabThera)

yes no

139 Was therapeutic rituximab (anti-CD20, Rituxan, Mab Thera) stopped?

yes no

140 Date therapeutic rituximab (anti-CD20, Rituxan, Mab Thera) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

141 Was any other monoclonal antibody given as therapy?

yes no

142 Was therapeutic other monoclonal antibody stopped?

yes no

143 Date therapeutic other monoclonal antibody stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

144 Specify other monoclonal antibody: _____

145 Was lenalidomide (Revlimid) given as therapy?

yes no

146 Was therapeutic lenalidomide (Revlimid) stopped?

yes no

147 Date therapeutic lenalidomide (Revlimid) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

148 Was mycophenolate mofetil (MMF, Cellcept) given as therapy?

yes no

149 Was therapeutic mycophenolate mofetil (MMF, Cellcept) stopped?

yes no

150 Date therapeutic mycophenolate mofetil (MMF, Cellcept) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

151 Was photopheresis / extracorporeal phototherapy (ECP)given as therapy?

yes no

152 Was therapeutic photopheresis / extracorporeal phototherapy (ECP) stopped?

yes no

153 Date therapeutic photopheresis / extracorporeal phototherapy (ECP) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

154 Was sirolimus (Rapamune) given as therapy?

yes no

155 Was therapeutic sirolimus (Rapamune) stopped?

yes no

156 Date therapeutic sirolimus (Rapamune) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

157 Was tacrolimus (FK506, Prograf) given as therapy?

yes no

158 Was therapeutic tacrolimus (FK506, Prograf) stopped?

yes no

159 Date therapeutic tacrolimus (FK506, Prograf) stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

160 Was thalidomide (Thalomid) given as therapy?

yes no

161 Was therapeutic thalidomide (Thalomid)stopped?

yes no

162 Date therapeutic thalidomide (Thalomid)stopped: __ __ __ __ - __ __- __ __ date estimated Date unknown

163 Was any other immunosuppressive drug given as therapy?

yes no

164 Was the other therapeutic immunosuppressive drug stopped?

yes no

165 Date other therapeutic immunosuppressive drug stopped: __ __ __ __ - __ __- __ __ 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)?

yes no

168 Specify other treatment(s): _____

Status of Hematologic Engraftment

Questions: 169 - 174

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?

predominantly or completely donor (>= 80% donor chimerism)

Mixed chimerism

only host T-cells detected (< 5% donor)

Unknown

170 Most recent date T-cell engraftment was assessed: __ __ __ __ - __ __- __ __ Date of most recent T-cell engraftment assessment unknown

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

predominantly or completely donor (>= 80% donor chimerism)

Mixed chimerism

only host B-cells detected (< 5% donor)

Unknown

172 Most recent date B-cell engraftment was assessed: __ __ __ __ - __ __- __ __ Date of most recent B-cell engraftment assessment unknown?

173 What is the current status of myeloid engraftment?

predominantly or completely donor (>= 80% donor chimerism)

Mixed chimerism

only host myeloid cells detected (< 5% donor)

Unknown

174 Most recent date myeloid engraftment was assessed: __ __ __ __ - __ __- __ __ Date of most recent myeloid engraftment assessment unknown

First Name: _____ Last Name: _____

Phone number: _____ Fax number: _____

E-mail address: _____