Form 2034 R3.0: X-Linked Lymphoproliferative Syndrome (XLP) Pre-HCT Data

Center: CRID:

Key Fields

Sequence Number: _____

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

CIBMTR Center Number: _____

CIBMTR Recipient ID: _____

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

HCT type (check all that apply):

- Autologous
- Allogeneic, unrelated
- Allogeneic, related

Product type (check all that apply):

- Bone marrow
- PBSC
- Single cord blood unit
- Multiple cord blood units
- Other product

Specify: _____

Subsequent Transplant

Is this the report of a second or subsequent transplant for the same disease?

yes no

Disease Assessment at Diagnosis

Questions: 1 - 23

1 Is this recipient a registered participant in the United States Immunodeficiency Network (USIDNET)?

yes no

2 USIDNET ID: _____

3 What was the date of diagnosis? __ __ __ __ - __ __- __ __

4 Was genetic testing used to confirm the diagnosis?

yes no Unknown

5 XLP1 / XLP (SH2D1A)

Yes No Unknown Not done

6 XLP2 / XIAP (BIRC4)

Yes No Unknown Not done

7 Other mutation

Yes No Unknown Not done

8 Specify other mutation: _____

9 Was documentation submitted to the CIBMTR?
(e.g. pathology report)

yes no

10 Was X-linked inheritance demonstrated in the recipient's maternal family members?

yes no Unknown

Specify if the following disorders were present at diagnosis:

11 Aplastic anemia

yes no

12 Colitis

yes no

13 Epstein-Barr Virus (EBV) infection with evidence of Hemophagocytic Lymphohistiocytosis (HLH)

yes no

14 EBV infection without HLH

yes no

15 Hypogammaglobulinemia

yes no

16 Lymphoproliferative disorder

yes no

17 Lymphoma

yes **- Also complete Form 2018 - LYM**

no

18 Psoriasis

yes no

19 Vasculitis

yes no

Specify the system(s) affected by vasculitis:

20 Central nervous system

yes no

21 Pulmonary system

yes no

22 Other vasculitis involvement

yes no

23 Specify other vasculitis involvement: _____

History of Epstein Barr Virus (EBV) Infection

Questions: 24 - 32

24 Is there a history of EBV infection?

Yes No Not evaluated

Specify results used for diagnosis of EBV:

25 In situ hybridization

Positive Negative Not Done

26 Polymerase chain reaction (PCR)

Positive Negative Not Done

27 Serology

Positive Negative Not Done

Specify results:

28 EBNA

Positive Negative

29 Early antigen

Positive Negative

30 Viral capsid IgG

Positive Negative

31 Viral capsid IgM

Positive Negative

32 Was documentation submitted to the CIBMTR?

yes no

Assessment of Immunologic Function at Diagnosis

Questions: 33 - 51

33 NK cell function

Absent (≤ 10% lower limit of normal)

Decreased (11-50% lower limit of normal)

Normal

Unknown

34 Invariant natural killer T-cells (iNKT)

Known Unknown

35 _____ m m³

36 Mucosal-associated invariant T-cells (MAIT)

Known Unknown

37 _____ m m³

38 Upper limit of normal of MAIT: _____ m m³

39 Lower limit of normal of MAIT: _____ m m³

40 Signaling lymphocyte activation molecule (SLAM)-associated protein (SAP) expression

Positive Negative Not Done

41 XIAP protein expression

Positive Negative Not Done

42 Did the recipient receive supplemental intravenous immunoglobulins (IVIG)?

yes no

43 Was therapy ongoing within three months of immunoglobulin testing?

yes no

Specify the quantitative immunoglobulins measured at diagnosis:

44 IgG

Known Unknown

45 _____ mg/dL g/dL g/L

46 IgM

Known Unknown

47 _____ mg/dL g/dL g/L

48 IgA

Known Unknown

49 _____ mg/dL g/dL g/L

50 IgE

Known Unknown

51 _____ IU/mL

Disease Assessment Between Diagnosis and the Start of the Preparative Regimen (Conditioning) Questions: 52 - 104

52 Was HLH present?

yes no

53 Was the HLH triggered by an acute EBV infection?

yes no Unknown

54 Was the HLH triggered by any other known condition(s)?

yes no Unknown

55 Bacterial infection

yes no

56 Fungal infection

yes no

57 Malignancy

yes no

58 Virus (not EBV)

yes no

Other Virus (1) Questions: 59 - 60

59 Specify virus _____

60 Specify other virus: _____

61 Other cause

yes no

62 Specify other cause: _____

Specify site(s) where HLH was present:

63 Bone marrow

yes no

64 Cerebrospinal fluid (CSF)

yes no

65 Liver

yes no

66 Lymph nodes

yes no

67 Spleen

yes no

68 Other site

yes no

69 Specify other site: _____

70 Was therapy given for HLH?

yes no

Line of Therapy (1) **Questions: 71 - 88**

Specify therapy given for HLH:

71 Date therapy started

Known Unknown

72 Date started: __ __ __ __ - __ __- __ __

73 Date therapy stopped

Known Unknown

74 Date stopped: __ __ __ __ - __ __- __ __

75 Alemtuzumab (Campath)

yes no

76 Specify total dose given: _____ mg

77 Antithymocyte globulin (ATG)

yes no

78 Corticosteroids
(intrathecal) (e.g. IT A-Hydrocort)

yes no

79 Corticosteroids
(systemic) (e.g., Dexamethasone)

yes no

80 Cyclosporine (CSA, Neoral, Sandimmune)

yes no

81 Etoposide (VP-16, VePesid)

yes no

82 Intrathecal methotrexate (IT MTX)

yes no

83 IVIG

yes no

84 Rituximab (Rituxan, MabThera)

yes no

85 Specify number of doses given: _____

86 Teniposide (VM26)

yes no

87 Other systemic therapy

yes no

88 Specify other systemic therapy: _____

89 Did colitis develop?

yes no Unknown

90 Did vasculitis develop?

yes no Unknown

Specify vasculitis involvement:

91 Central nervous system

yes no

92 Pulmonary system

yes no

93 Other vasculitis involvement

yes no

94 Specify other vasculitis involvement: _____

95 Did the recipient develop lymphoma?

yes **– Also complete Form 2018 - LYM**

no

96 Was the lymphoma associated with an EBV infection?

yes no Unknown

97 Is the tumor EBV positive?

yes no Unknown

98 Was documentation submitted to the CIBMTR?
(e.g. pathology report)

yes no

99 Did the recipient develop hypogammaglobulinemia?

yes no

100 Did the recipient develop aplastic anemia?

yes no

Specify therapy given for aplastic anemia:

101 Growth factor

yes no

102 Immunosuppression

yes no

103 Other therapy

yes no

104 Specify other therapy: _____

Disease Status at Last Evaluation Prior to the Start of the Preparative Regimen (Conditioning)

Questions: 105 - 130

105 Specify the status of HLH

- Active
- Inactive (quiescent)
- Not applicable

106 Was colitis active?

Yes No Unknown Not applicable

107 Was the recipient receiving therapy for colitis?

yes no Unknown

108 Was the CNS vasculitis active?

Yes No Unknown Not applicable

109 Was the recipient receiving therapy for CNS vasculitis?

yes no Unknown

110 Was pulmonary vasculitis active?

Yes No Unknown Not applicable

111 Was the recipient receiving therapy for pulmonary vasculitis?

yes no Unknown

112 Was the other vasculitis active?

Yes No Unknown Not applicable

113 Was the recipient receiving therapy for other vasculitis?

yes no Unknown

Specify the clinical and laboratory features assessed at last evaluation prior to the preparative regimen:

114 Serum ferritin

Known Unknown

115 _____ µg/L

116 Date sample collected: __ __ __ __ - __ __- __ __

117 Soluble interleukin-2 receptor (sIL-2R)

Known Unknown

118 _____ mg/dL µmol/L U/mL

119 Date sample collected: __ __ __ __ - __ __- __ __

120 Triglycerides

Known Unknown

121 _____ mg/dL mmol/L

122 Date sample collected: __ __ __ __ - __ __- __ __

123 Fibrinogen antigen assay (factor I; fibrinogen activity; functional fibrinogen; fibrinogen antigen)

Known Unknown

124 _____ g/dL mg/dL µmol/L g/L

125 Date sample collected: __ __ __ __ - __ __- __ __

126 Bone marrow aspirate / biopsy evidence of hemophagocytosis

Present Absent Not done

Specify the cerebrospinal fluid findings:

127 Protein

Normal Elevated Not done

128 WBC count

Normal (≤ 5 cells/μL)

Elevated (> 5 cells/μL)

Not done

129 Was donor testing for XLP done prior to HCT?

Yes

No

Not applicable (related female and/or unrelated donor)

130 Was there evidence of XLP?

yes no

First Name: _____

Last Name: _____

E-mail address: _____

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