Form 2039 R3.0: Hemophagocytic Lymphohistiocytosis (HLH) 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 - 22

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 Is there a family history of hemophagocytic disorders?

yes no Unknown

Specify affected member(s):

5 Aunt(s)

yes no

6 Uncle(s)

yes no

7 Cousin(s)

yes no

8 Sibling(s)

yes no

9 Other family member(s)

yes no

10 Specify relationship: _____

11 Is there a family history of consanguinity (inter-familial marriage / descent from common ancestors)?

yes no Unknown

12 Was genetic testing used to confirm the diagnosis?

yes no Unknown

Specify genetic mutation(s) identified:

13 FHL 2 / Perforin deficiency (PRF1)

Yes No Unknown Not done

14 FHL 3 / MUNC 13-4 (UNC13D)

Yes No Unknown Not done

15 FHL 4 / Syntaxin 11 (STX11)

Yes No Unknown Not done

16 FHL 5 / Munc 18-2 (STXBP2)

Yes No Unknown Not done

17 IL2-inducible T-cell kinase (ITK)

Yes No Unknown Not done

18 Other mutation

Yes No Unknown Not done

19 Specify other mutation: _____

20 Were central nervous system (CNS) abnormalities found on computed tomography (CT or CAT) or magnetic resonance imaging (MRI) scans?

yes no Unknown

21 Date scan was performed: __ __ __ __ - __ __- __ __

22 Was documentation submitted to the CIBMTR?
(e.g. CT or MRI scan report)

yes no

Clinical Features and Laboratory Studies at Diagnosis

Questions: 23 - 47

23 Anemia
(Hgb < 9 g/dL)

yes no Unknown

24 Degranulation assay of NK cells
(as defined by local laboratory)

Normal Abnormal Unknown

25 Fevers
(>38.5° C or > 101.3° F for >7 days within 1 week of diagnosis)

yes no Unknown

26 Hepatomegaly (liver edge palpable > 3 cm below right costal margin)

yes no Unknown

27 Serum ferritin

Known Unknown

28 _____ ng/mL(µg/L)

29 Triglycerides

Known Unknown

30 _____ mg/dL mmol/L

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

Known Unknown

32 _____ g/dL mg/dL µmol/L g/L

33 NK cell function

Absent (≤10% lower limit of normal)

Decreased (11-50% lower limit of normal)

Normal

Unknown

34 Neutropenia (ANC < 1.0 x 10^9 /L)

yes no Unknown

35 Soluble interleukin-2 receptor alpha chain (sCD25)
(as defined by local laboratory)

Normal Elevated Unknown

36 Splenomegaly (spleen palpable > 3 cm below left costal margin)

yes no Unknown

37 Thrombocytopenia (platelets < 100 x 10^9 /L)

yes no Unknown

Specify the cerebrospinal fluid findings at diagnosis

38 Neopterin level

Normal Elevated Not done

39 Protein

Normal Elevated Not done

40 WBC count

Normal (≤ 5 cells/µL)

Elevated (> 5 cells/µL)

Not done

Specify the site(s) where hemophagocytosis was documented at diagnosis:

41 Bone marrow

yes no

42 Cerebrospinal fluid (CSF)

yes no

43 Liver

yes no

44 Lymph nodes

yes no

45 Spleen

yes no

46 Other site

yes no

47 Specify other site: _____

Disease Assessment Between Diagnosis and the Start of the Preparative Regimen (Conditioning) Questions: 48 - 58

48 Were central nervous system (CNS) abnormalities found on computed tomography (CT or CAT) or magnetic resonance imaging (MRI) scans?

yes no Unknown

49 Date scan was performed: __ __ __ __ - __ __- __ __

50 Were there any clinical neurologic abnormalities present?

yes no Unknown

Specify neurologic abnormalities:

51 Abnormal gait

yes no

52 Cranial nerve palsies

yes no

53 Developmental delay

yes no

54 Motor weakness

yes no

55 Seizures

yes no

56 Sensory deficits

yes no

57 Other neurologic abnormality

yes no

58 Specify other neurologic abnormality: _____

History of Infection at Any Time Prior to the Preparative Regimen (Conditioning)

Questions: 59 - 74

Specify documented infection(s) associated with HLH:

59 Was an infection documented?

yes no

Specify infection(s):

60 Cytomegalovirus (CMV)

yes no

61 Specify the test method used for diagnosis of CMV
(check only one)

Antigen

Polymerase chain reaction (PCR)

Shell vial test

62 Epstein-Barr virus (EBV)

yes no

Specify results used for diagnosis of EBV infection:

63 In situ hybridization

Positive Negative Not Done

64 Polymerase chain reaction (PCR)

Positive Negative Not Done

65 Serology

Positive Negative Not Done

Specify titers:

66 EBNA

Positive Negative

67 Early antigen

Positive Negative

68 Viral capsid IgG

Positive Negative

69 Viral capsid IgM

Positive Negative

70 Was documentation submitted to the CIBMTR?

yes no

71 Other infection

yes no

Other Infection (1)

Questions: 72 - 74

72 Specify other infection

_____

73 If other organism, specify: _____

74 Site

_____

Pre-HCT Therapy

Questions: 75 - 107

75 Was therapy given?

yes no

Line of Therapy (1)

Questions: 76 - 107

Line of Therapy

76 Specify the purpose of therapy

Induction

Maintenance

Treatment for disease relapse / reactivation

77 Date therapy started

Known Unknown

78 Date started: __ __ __ __ - __ __- __ __

79 Alemtuzumab (Campath)

yes no

80 Specify total dose given: _____ mg

81 Antithymocyte globulin (ATG)

yes no

82 Corticosteroids
(intrathecal) (e.g., IT-A-Hydrocort)

yes no

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

yes no

84 Cyclosporine (CSA, Neoral, Sandimmune)

yes no

85 Etoposide (VP-16, VePesid)

yes no

86 Intrathecal methotrexate (IT MTX)

yes no

87 IVIG

yes no

88 Teniposide (VM26)

yes no

89 Other therapy

yes no

90 Specify other therapy: _____

91 Was this therapy given following the HLH-94 / HLH 2004 protocol of the Histiocyte Society?

yes no Unknown

Specify response to therapy:

92 Was CNS disease inactive?

yes no Unknown

93 Normal or stable CT or MRI of CNS

yes no Unknown

94 Neopterin level

Normal Elevated Not done

95 Protein

Normal Elevated Not done

96 WBC count

Normal (≤ 5 cells/μL)

Elevated (> 5 cells/μL)

Not done

97 Was systemic disease inactive?

yes no Unknown

98 ANC > 1.0 x 10^9 /L (without growth factor support)

yes no Unknown

99 Hemoglobin ≥9 g/dL without transfusion

yes no Unknown

100 Hepatomegaly resolved (≤3cm below costal margin)

yes no Unknown

101 Normal fibrinogen

yes no Unknown

102 Normal triglycerides

yes no Unknown

103 Platelets > 100 x 10^9 /L without transfusion

yes no Unknown

104 Splenomegaly resolved (≤3 cm below costal margin)

yes no Unknown

105 Were there any signs of disease relapse / reactivation?

yes no

106 Specify the date of the relapse / reactivation: __ __ __ __ - __ __- __ __

107 Specify the site of the relapse / reactivation

CNS Systemic CNS and systemic

Clinical Features and Laboratory Studies Prior to the Preparative Regimen (Conditioning)

Questions: 108 - 127

108 Anemia
(Hgb < 9 g/dL)

yes no Unknown

109 Degranulation assay of NK cells
(as defined by local laboratory)

Normal Abnormal Unknown

110 Fevers
(>38.5° C or > 101.3° F for > 7 days)

yes no Unknown

111 Hepatomegaly (liver edge palpable > 3 cm below right costal margin)

yes no Unknown

112 Serum ferritin

Known Unknown

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Unknown

119 Neutropenia (ANC < 1.0 x 10^9 /L)

yes no Unknown

120 Soluble interleukin-2 receptor alpha chain (sCD25)
(as defined by local laboratory)

Normal Elevated Unknown

121 Splenomegaly (spleen palpable > 3 cm below left costal margin)

yes no Unknown

122 Thrombocytopenia (platelets < 100 x 10^9 /L)

yes no Unknown

Specify the cerebrospinal fluid findings at last assessment prior to the start of the preparative regimen

123 Neopterin level

Normal Elevated Not done

124 Protein

Normal Elevated Not done

125 WBC count

Normal (≤ 5 cells/µL)

Elevated (> 5 cells/µL)

Not done

126 Were central nervous system (CNS) abnormalities found on computed tomography (CT or CAT) or magnetic resonance imaging (MRI) scans?

yes no Unknown

127 Date scan was performed: __ __ __ __ - __ __- __ __

First Name: _____

Last Name: _____

E-mail address: _____

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