Form 2134 R3.0: X-Linked Lymphoproliferative Syndrome (XLP) Post-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: _____

Visit

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

Specify: _____

Disease Assessment Since the Date of Last Report

Questions: 1 - 19

1 Did the recipient have lymphoma at the time of HCT?

yes no

2 Did the recipient develop lymphoma or have persistent disease since the date of last report?

yes no

3 Specify current status of lymphoma

- Complete remission (CR) -complete disappearance of all known disease for ≥ 4 weeks
- Partial remission (PR) - ≥50% reductions in greatest diameter of all sites of known disease and no new sites
- Stable disease (SD) - <50% reductions in greatest diameter of all sites of known disease
- Progressive disease (PD) - increase in size of known disease, or new sites of disease
- Unknown
- Not assessed

4 Did colitis persist or develop since the date of the last report?

yes no Unknown

5 What is the status of colitis?

Active Inactive Unknown

6 Was the recipient receiving therapy for colitis?

yes no Unknown

7 Did vasculitis persist or develop since the date of the last report?

yes no Unknown

Specify vasculitis involvement:

8 Central nervous system

yes no

9 What is the status of the CNS vasculitis?

Active Inactive Unknown

10 Was the recipient receiving therapy for CNS vasculitis?

yes no Unknown

11 Pulmonary system

yes no

12 What is the status of the pulmonary vasculitis?

Active Inactive Unknown

13 Was the recipient receiving therapy for pulmonary vasculitis?

yes no Unknown

14 Other vasculitis involvement

yes no

15 Specify other vasculitis involvement: _____

16 What is the status of other vasculitis?

Active Inactive Unknown

17 Was the recipient receiving therapy for other vasculitis?

yes no Unknown

18 Did the recipient have hemophagocytic lymphohistiocytosis (HLH) prior to transplant or did it present since the date of last report?

yes no

19 Specify the status of the HLH disease since the date of the last report:

Active

Inactive (quiescent)

Current Assessment of Immunologic Function Post-HCT

Questions: 20 - 30

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

yes no

21 Was therapy ongoing within three months of immunoglobulin testing?

yes no

22 IgG

Known Unknown

23 _____ mg/dL g/dL g/L

24 IgM

Known Unknown

25 _____ mg/dL g/dL g/L

26 IgA

Known Unknown

27 _____ mg/dL g/dL g/L

28 IgE

Known Unknown

29 _____ IU/mL

30 NK cell function

Absent (≤ 10% lower limit of normal)

Decreased (11-50% lower limit of normal)

Normal

Unknown

Laboratory Studies at the Time of Evaluation for This Reporting Period

Questions: 31 - 41

31 Serum ferritin

Known Unknown

32 _____ µg/L

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

Known Unknown

34 _____ mg/dL µmol/L U/mL

35 Triglycerides

Known Unknown

36 _____ mg/dL mmol/L

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

Known Unknown

38 _____ g/dL mg/dL µmol/L g/L

39 Bone marrow aspirate / biopsy evidence of hemophagocytosis

Present Absent Not done

Specify the cerebrospinal fluid findings:

40 Protein

Normal Elevated Not done

41 WBC count

Normal Elevated Not done

First Name: _____

Last Name: _____

E-mail address: _____

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