### Key Fields

| Sequence Number: | ____________________________ |

ELSE GOTO Date Received:

| Date Received: | __________ YYYY - MM - DD |

ELSE GOTO CIBMTR Center Number:

| CIBMTR Center Number: | ____________________________ |

ELSE GOTO CIBMTR Recipient ID:

| CIBMTR Recipient ID: | ____________________________ |

ELSE GOTO Today's Date:

| Today's Date: | __________ YYYY - MM - DD |

ELSE GOTO Date of HSCT for which this form is being completed:

| Date of HSCT for which this form is being completed: | __________ YYYY - MM - DD |

ELSE GOTO Autologous

**HSCT type:** (check all that apply)

- [ ] Autologous

ELSE GOTO allogeneic unrelated

- [ ] allogeneic unrelated

ELSE GOTO syngeneic(identical twin)

ELSE GOTO Marrow

**Product type:** (check all that apply)

- [ ] Marrow

ELSE GOTO PBSC

- [ ] PBSC

ELSE GOTO Cord blood

- [ ] Cord blood

ELSE GOTO Other product
☐ Other product
IF Other product := EXISTS
THEN GOTO Specify:
ELSE GOTO If this is a report of a second or subsequent transplant, check here and continue with question 43.

Specify:
ELSE GOTO If this is a report of a second or subsequent transplant, check here and continue with question 43.

☐ If this is a report of a second or subsequent transplant, check here and continue with question 43.
IF This is a report of a second or subsequent transplant := checked
THEN GOTO (43) Did the recipient receive IVIG within two months prior to the above immunoglobulin measurement?
ELSE GOTO (1) What was the date of diagnosis of X-Linked Lymphoproliferative Syndrome?

X-Linked Lymphoproliferative Syndrome Pre-HSCT Data Questions: 1-60

This form must be accompanied by Form 2000-Recipient Baseline Data. All information in the box about, including the date, should be identical with the corresponding Form 2000. Information should come from an actual examination by the Transplant Center physician or the physician who is following the recipient pre-HSCT or abstraction of the recipient's medical records.

1 What was the date of diagnosis of X-Linked Lymphoproliferative Syndrome? __ YYYY __ MM __ DD __

ELSE GOTO (2) Was the XLP diagnosis confirmed by genetic testing?

2 Was the XLP diagnosis confirmed by genetic testing?
   O yes
   O no
   O unknown
IF (2) Was the XLP diagnosis confirmed by genetic testing := yes
THEN GOTO (3) Is a copy of the diagnosis report attached?
ELSE GOTO (4) Aplastic Anemia

3 Is a copy of the diagnosis report attached?
   O yes
   O no
ELSE GOTO (4) Aplastic Anemia

Specify if the recipient displayed evidence of the following disorders at diagnosis of XLP:

4 Aplastic Anemia
   O yes
   O no
ELSE GOTO (5) Hemophagocytic disorder(fulminant infectious mononucleosis)

5 Hemophagocytic disorder(fulminant infectious mononucleosis)
   O yes
   O no
ELSE GOTO (6) Hypogammaglobulinemia

6 Hypogammaglobulinemia
   O yes
   O no
ELSE GOTO (7) Lymphoproliferative disorder

7 Lymphoproliferative disorder
   O yes
   O no
ELSE GOTO (8) Was X-linked inheritance demonstrated in the recipient's maternal family members?

8 Was X-linked inheritance demonstrated in the recipient's maternal family members?
   O yes
   O no
   O unknown
ELSE GOTO (9) Were pre-HSCT Epstein-Barr virus (EBV) serology titers determined?
History of Infection at Diagnosis

9 Were pre-HSCT Epstein-Barr virus (EBV) serology titers determined?
   O yes
   O no
   O unknown

IF (9) Were pre-HSCT Epstein-Barr virus (EBV) serology titers determined?:= yes
THEN GOTO (10) Date tested:
ELSE GOTO (15) Was hemophagocytic disorder (fulminant infectious mononucleosis) present at any time?

10 Date tested: __ YYMMDD

ELSE GOTO (11) Viral capsid IgG titer

   Specify titers tested:

11 Viral capsid IgG titer:
   O positive
   O negative
   O not tested

ELSE GOTO (12) Viral capsid IgM titer

12 Viral capsid IgM titer:
   O positive
   O negative
   O not tested

ELSE GOTO (13) Early antigen titer

13 Early antigen titer:
   O positive
   O negative
   O not tested

ELSE GOTO (14) EBNA titer

14 EBNA titer:
   O positive
   O negative
   O not tested

ELSE GOTO (15) Was hemophagocytic disorder (fulminant infectious mononucleosis) present at any time?

15 Was hemophagocytic disorder (fulminant infectious mononucleosis) present at any time?
   O yes
   O no

IF (15) Was hemophagocytic disorder (fulminant infectious mononucleosis) present at any time?:= yes
THEN GOTO (16) Bone marrow
ELSE GOTO (36) Was immunologic function tested at diagnosis?

   Specify site(s) of hemophagocytosis:

16 Bone marrow:
   O yes
   O no

ELSE GOTO (17) Cerebrospinal fluid (CSF)

17 Cerebrospinal fluid (CSF):
   O yes
   O no

ELSE GOTO (18) Liver

18 Liver:
   O yes
   O no

ELSE GOTO (19) Lymph nodes
19. Lymph nodes
   - yes
   - no
   ELSE GOTO (20) Spleen

20. Spleen
   - yes
   - no
   ELSE GOTO (21) Other site:

21. Other site:
   - yes
   - no
   IF (21) Other site:: = yes
   THEN GOTO (22) Specify other site:
   ELSE GOTO (23) Cyclosporine

22. Specify other site: __________________________
   ELSE GOTO (23) Cyclosporine

23. Cyclosporine
   - yes
   - no
   ELSE GOTO (24) Intrathecal methotrexate

24. Intrathecal methotrexate
   - yes
   - no
   ELSE GOTO (25) IVIG

25. IVIG
   - yes
   - no
   ELSE GOTO (26) Radiation therapy

26. Radiation therapy
   - yes
   - no
   IF (26) Radiation therapy:: = yes
   THEN GOTO (27) Specify radiation field:
   ELSE GOTO (29) Steriods

27. Specify radiation field: __________________________
   ELSE GOTO (28) radiation dose

28. Specify total dose: __________________________ cGy
   ELSE GOTO (29) Steriods

29. Steriods
   - yes
   - no
   ELSE GOTO (30) VM-16/VM-26

30. VM-16/VM-26
   - yes
   - no
   ELSE GOTO (31) Other drug
31 Other drug
  o yes
  o no
  IF (31) Other drug := yes
  THEN GOTO (32) Specify other drug:
  ELSE GOTO (33) Other treatment

32 Specify other drug: ____________________________
  ELSE GOTO (33) Other treatment

33 Other treatment
  o yes
  o no
  IF (33) Other treatment := yes
  THEN GOTO (34) Specify other treatment
  ELSE GOTO (35) Was the hemophagocytic syndrome triggered by an acute EBV infection?

34 Specify ____________________________
  ELSE GOTO (35) Was the hemophagocytic syndrome triggered by an acute EBV infection?

35 Was the hemophagocytic syndrome triggered by an acute EBV infection?
  o yes
  o no
  o unknown
  ELSE GOTO (36) Was immunologic function tested at diagnosis?

36 Was immunologic function tested at diagnosis?
  o yes
  o no
  o unknown
  IF (36) Was immunologic function tested at diagnosis? := yes
  THEN GOTO (37) Natural Killer cell function (specific cytosis of NK-sensitive target cell)
  ELSE GOTO (43) Did the recipient receive IVIG within two months prior to the above immunoglobulin measurement?

Specify findings at diagnosis:

37 Natural Killer cell function (specific cytosis of NK-sensitive target cell)
  o absent (<= 10% normal response)
  o decreased (11-50% normal response)
  o normal
  o increased
  o Not available
  ELSE GOTO (38) IgG prior to receiving IVIG

38 IgG prior to receiving IVIG
  o absent (<= 10% normal response)
  o decreased (11-50% normal response)
  o normal
  o increased
  o Not available
  ELSE GOTO (39) IgM prior to receiving IVIG

39 IgM prior to receiving IVIG
  o absent (<= 10% normal response)
  o decreased (11-50% normal response)
  o normal
  o increased
  o Not available
  ELSE GOTO (40) IgA prior to receiving IVIG
40 IgA prior to receiving IVIG
   O absent (<= 10% normal response)
   O decreased (11-50% normal response)
   O normal
   O increased
   O Not available
ELSE GOTO (41) IgE prior to receiving IVIG

41 IgE prior to receiving IVIG
   O absent (<= 10% normal response)
   O decreased (11-50% normal response)
   O normal
   O increased
   O Not available
ELSE GOTO (42) Is a copy of the immunologic report or other documentation attached?

42 Is a copy of the immunologic report or other documentation attached?
   O yes
   O no
ELSE GOTO (43) Did the recipient receive IVIG within two months prior to the above immunoglobulin measurement?

43 Did the recipient receive IVIG within two months prior to the above immunoglobulin measurement?
   O yes
   O no
   O unknown
ELSE GOTO (44) Did the recipient develop lymphoma prior to the preparative regimen?

44 Did the recipient develop lymphoma prior to the preparative regimen?
   O yes
   O no
IF (44) Did the recipient develop lymphoma prior to the preparative regimen?:
   THEN GOTO (45) Chemotherapy
   ELSE GOTO (50) Did the recipient develop hypogammaglobulinemia prior to the preparative regimen?

45 Specify treatment(s) given for lymphoma:
   Chemotherapy
   O yes
   O no
ELSE GOTO (46) Radiation

46 Radiation
   O yes
   O no
ELSE GOTO (47) What was the response of the lymphoma to treatment prior to the preparative regimen?

47 What was the response of the lymphoma to treatment prior to the preparative regimen?
   O complete response (CR)
   O partial response (PR)
   O progressive disease
   O not applicable/no treatment given
ELSE GOTO (48) Was the lymphoma associated with an EBV infection?

48 Was the lymphoma associated with an EBV infection?
   O yes
   O no
   O unknown
ELSE GOTO (49) Is a copy of the pathology report or other documentation attached?

49 Is a copy of the pathology report or other documentation attached?
   O yes
   O no
ELSE GOTO (50) Did the recipient develop hypogammaglobulinemia prior to the preparative regimen?
50 Did the recipient develop hypogammaglobulinemia prior to the preparative regimen?
   O yes
   O no
   IF (50) Did the recipient develop hypogammaglobulinemia prior to the preparative regimen?:= yes
      THEN GOTO (51) IVIG
      ELSE GOTO (54) Did recipient develop aplastic anemia prior to the preparative regimen?
         Specify treatment(s) given for hypogammaglobulinemia
   51 IVIG
      O yes
      O no
      ELSE GOTO (52) Other treatment
   52 Other treatment
      O yes
      O no
      IF (52) Other treatment:= yes
         THEN GOTO (53) Specify:
         ELSE GOTO (54) Did recipient develop aplastic anemia prior to the preparative regimen?
   53 Specify:
      ELSE GOTO (54) Did recipient develop aplastic anemia prior to the preparative regimen?
   54 Did recipient develop aplastic anemia prior to the preparative regimen?
      O yes
      O no
      IF (54) Did recipient develop aplastic anemia prior to the preparative regimen?:= yes
      THEN GOTO (55) Growth factor
      ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?
         Specify treatment(s) given for aplastic anemia:
   55 Growth factor
      O yes
      O no
      ELSE GOTO (56) Immunosuppression
   56 Immunosuppression
      O yes
      O no
      ELSE GOTO (57) Other treatment
   57 Other treatment
      O yes
      O no
      IF (57) Other treatment:= yes
      THEN GOTO (58) Specify:
      ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?
         Specify:
   58 Specify:
      ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?
   59 Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?
      O yes
      O no
      O unknown
      IF (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?:= yes
      THEN GOTO (60) Is a copy of the MRI report attached?
      ELSE GOTO First name
| 60 Is a copy of the MRI report attached? 
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ELSE GOTO First name

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ELSE GOTO Last name

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ELSE GOTO E-mail address:

ELSE GOTO End of Form