# 2034: X-Linked Lymphoproliferative Syndrome

## Pre-HSCT Data

**Registry Use Only**

### Key Fields

<table>
<thead>
<tr>
<th>Sequence Number:</th>
<th>ELSE GOTO Date Received:</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Date Received: <strong>YYYY</strong> <strong>MM</strong> <strong>DD</strong></td>
</tr>
<tr>
<td>CIBMTR Center Number:</td>
<td>ELSE GOTO CIBMTR Center Number:</td>
</tr>
<tr>
<td></td>
<td>CIBMTR Recipient ID:</td>
</tr>
<tr>
<td></td>
<td>ELSE GOTO CIBMTR Recipient ID:</td>
</tr>
<tr>
<td></td>
<td>Today’s Date: <strong>YYYY</strong> <strong>MM</strong> <strong>DD</strong></td>
</tr>
<tr>
<td></td>
<td>ELSE GOTO Date of HSCT for which this form is being completed:</td>
</tr>
<tr>
<td></td>
<td>Date of HSCT for which this form is being completed: <strong>YYYY</strong> <strong>MM</strong> <strong>DD</strong></td>
</tr>
<tr>
<td>Autologous</td>
<td>ELSE GOTO Autologous</td>
</tr>
<tr>
<td>HSCT type: (check all that apply)</td>
<td></td>
</tr>
<tr>
<td>☐ Autologous</td>
<td>ELSE GOTO allogeneic unrelated</td>
</tr>
<tr>
<td>☐ allogeneic unrelated</td>
<td>ELSE GOTO allogeneic related:</td>
</tr>
<tr>
<td>☐ allogeneic related</td>
<td>ELSE GOTO syngeneic(identical twin)</td>
</tr>
</tbody>
</table>

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Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
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☐ 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 If this is a report of a second or subsequent transplant, check here and continue with question 43.= 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

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? ___________ ___________ ___________ MM DD

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

☐ yes
☐ no
☐ 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?
☐ yes

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CIBMTR Center Number: ___________________  CIBMTR Recipient ID: ___________________

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?

IF (9) Were pre-HSCT Epstein-Barr virus (EBV) serology titers determined?: = yes

ELSE GOTO (10) Date tested:

10 Date tested: ____________ YYYY ____________ MM ____________ DD

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

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

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

   Specify therapy given for hemophagocytosis:

23 Cyclosporine
   O yes
   O no
   ELSE GOTO (24) Intrathecal methotrexate

24 Intrathecal methotrexate
   O yes
   O no
   ELSE GOTO (25) IVIG

25 IVIG
   O yes
   O no
   ELSE GOTO (26) Radiation therapy

26 Radiation therapy
   O yes
   O 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
   O yes
   O no
   ELSE GOTO (30) VM-16/VM-26
CIBMTR Center Number: ___ ___ ___ ___ ___  
CIBMTR Recipient ID: ___ ___ ___ ___ ___ ___ ___ ___ ___ ___ ___ ___ ___ 

Today's Date:  
Month  Day  20  Year  
Infusion Date:  
Month  Day  20  Year  
CIBMTR Center Number: 

CIBMTR Center Number: ___________________
CIBMTR Recipient ID: ___________________

30 VM-16/VM-26
  O yes  
  O 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?

37 Natural Killer cell function(specific cytosis of NK-sensitive target cell)
  O absent (<= 10% normal response)  
  O decreased(11-50% normal response)
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CIBMTR Recipient ID: ___ ___ ___ ___ ___ ___ ___ ___ ___

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<tr>
<th>CIBMTR Center Number: __________</th>
<th>CIBMTR Recipient ID: __________</th>
</tr>
</thead>
<tbody>
<tr>
<td>51 IVIG</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>ELSE GOTO (52) Other treatment</td>
<td></td>
</tr>
<tr>
<td>52 Other treatment</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>IF (52) Other treatment:= yes</td>
<td></td>
</tr>
<tr>
<td>THEN GOTO (53) Specify:</td>
<td></td>
</tr>
<tr>
<td>ELSE GOTO (54) Did recipient develop aplastic anemia prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>53 Specify:</td>
<td></td>
</tr>
<tr>
<td>ELSE GOTO (54) Did recipient develop aplastic anemia prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>54 Did recipient develop aplastic anemia prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>IF (54) Did recipient develop aplastic anemia prior to the preparative regimen?:= yes</td>
<td></td>
</tr>
<tr>
<td>THEN GOTO (55) Growth factor</td>
<td></td>
</tr>
<tr>
<td>ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>Specify treatment(s) given for aplastic anemia:</td>
<td></td>
</tr>
<tr>
<td>55 Growth factor</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>ELSE GOTO (56) Immunosuppression</td>
<td></td>
</tr>
<tr>
<td>56 Immunosuppression</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>ELSE GOTO (57) Other treatment</td>
<td></td>
</tr>
<tr>
<td>57 Other treatment</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
<tr>
<td>IF (57) Other treatment:= yes</td>
<td></td>
</tr>
<tr>
<td>THEN GOTO (58) Specify:</td>
<td></td>
</tr>
<tr>
<td>ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>Specify:</td>
<td></td>
</tr>
<tr>
<td>ELSE GOTO (59) Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>59 Did the recipient have magnetic resonance imaging(MRI) of the brain immediately prior to the preparative regimen?</td>
<td></td>
</tr>
<tr>
<td>O yes</td>
<td>O no</td>
</tr>
</tbody>
</table>
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?
  O yes
  O no
ELSE GOTO First name

First Name: __________________________
ELSE GOTO Last name

Last Name: __________________________
ELSE GOTO Phone number:

Phone number: ________________________
ELSE GOTO Fax number:

Fax number: __________________________
ELSE GOTO E-mail address:

E-mail address: ________________________
ELSE GOTO End of Form