



X-Linked Lymphoproliferative Syndrome Pre-HSCT Data

Registry Use Only

Sequence Number:

Date Received:

CIBMTR Center Number:

CIBMTR Recipient ID:

Today's Date: / / (20)

Date of HSCT for which this form is being completed: / / (20)

HSCT type: autologous allogeneic, unrelated allogeneic, related syngeneic (identical twin)

Product type: marrow PBSC cord blood other product, specify: _____

This form must be accompanied by Form 2000 – Recipient Baseline Data. All information in the box above, 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.

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

1. What was the date of diagnosis of X-Linked Lymphoproliferative Syndrome? / /

2. Was the XLP diagnosis confirmed by genetic testing?

- 1 yes
- 2 no
- 3 unknown

3. Is a copy of the diagnosis report attached?

- 1 yes
- 2 no

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

- 4. 1 yes 2 no Aplastic anemia
- 5. 1 yes 2 no Hemophagocytic disorder (fulminant infectious mononucleosis)
- 6. 1 yes 2 no Hypogammaglobulinemia
- 7. 1 yes 2 no Lymphoproliferative disorder

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

- 1 yes
- 2 no
- 3 unknown

History of Infection at Diagnosis

9. Were pre-HSCT Epstein-Barr virus (EBV) serology titers determined?

- 1 yes
- 2 no
- 3 unknown

10. Date tested: / / (20)

Specify titers tested:

- 11. 1 positive 2 negative 3 not tested Viral capsid IgG titer
- 12. 1 positive 2 negative 3 not tested Viral capsid IgM titer
- 13. 1 positive 2 negative 3 not tested Early antigen titer
- 14. 1 positive 2 negative 3 not tested EBNA titer

CIBMTR Center Number:

CIBMTR Recipient ID:

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

- 1 yes
- 2 no

Specify site(s) of hemophagocytosis:

- 16. 1 yes 2 no Bone marrow
- 17. 1 yes 2 no Cerebrospinal fluid (CSF)
- 18. 1 yes 2 no Liver
- 19. 1 yes 2 no Lymph nodes
- 20. 1 yes 2 no Spleen
- 21. 1 yes 2 no Other site

22. Specify other site:

Specify therapy given for hemophagocytosis:

- 23. 1 yes 2 no Cyclosporine
- 24. 1 yes 2 no Intrathecal methotrexate
- 25. 1 yes 2 no IVIG
- 26. 1 yes 2 no Radiation therapy
- 29. 1 yes 2 no Steroids
- 30. 1 yes 2 no VP-16 / VM-26
- 31. 1 yes 2 no Other drug

27. Specify radiation field:

28. Specify total dose: cGy

32. Specify other drug:

34. Specify:

35. Was the hemophagocytic syndrome triggered by an acute EBV infection?

- 1 yes
- 2 no
- 3 unknown

36. Was immunologic function tested at diagnosis?

- 1 yes
- 2 no
- 3 unknown

Specify findings at diagnosis:

	Absent (≤ 10% normal)	Decreased (11–50% normal)	Normal	Increased	Not available
37. Natural killer cell function (specific cytotoxicity of NK-sensitive target cell)	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
38. IgG prior to receiving IVIG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
39. IgM prior to receiving IVIG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
40. IgA prior to receiving IVIG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>
41. IgE prior to receiving IVIG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>

42. Is a copy of the immunologic report or other documentation attached?

- 1 yes
- 2 no

43. Did the recipient receive IVIG within two months prior to the above immunoglobulin measurement?

- 1 yes
- 2 no
- 3 unknown

CIBMTR Center Number:

CIBMTR Recipient ID:

44. Did the recipient develop lymphoma prior to the preparative regimen?

- 1 yes
- 2 no

Specify treatment(s) given for lymphoma:

45. 1 yes 2 no Chemotherapy

46. 1 yes 2 no Radiation

47. What was the response of the lymphoma to treatment prior to the preparative regimen?

- 1 complete response
- 2 partial response
- 3 progressive disease
- 4 not applicable / no treatment given

48. Was the lymphoma associated with an EBV infection?

- 1 yes
- 2 no
- 3 unknown

49. Is a copy of the pathology report or other documentation attached?

- 1 yes
- 2 no

50. Did the recipient develop hypogammaglobulinemia prior to the preparative regimen?

- 1 yes
- 2 no

Specify treatment(s) given for hypogammaglobulinemia:

51. 1 yes 2 no IVIG

52. 1 yes 2 no Other treatment → 53. Specify:

54. Did the recipient develop aplastic anemia prior to the preparative regimen?

- 1 yes
- 2 no

Specify treatment(s) given for aplastic anemia:

55. 1 yes 2 no Growth factor

56. 1 yes 2 no Immunosuppression

57. 1 yes 2 no Other treatment → 58. Specify:

59. Did the recipient have magnetic resonance imaging (MRI) of the brain immediately prior to the preparative regimen?

- 1 yes
- 2 no
- 3 unknown

60. Is a copy of the MRI report attached?

- 1 yes
- 2 no

61. Signed: _____

Person completing form

Please print name: _____

Phone: (_____) _____

Fax: (_____) _____

E-mail address: _____