CIBMTR®
CENTER FOR INTERNATIONAL BLOOD & MARROW TRANSPLANT RESEARCH

CIBMTR Center Number: | | | | | |

CIBMTR Recipient ID: | | | | | | | | | | |

Today's Date: Month | | Day | | | 2 | 0 | | | Year

Date of HSCT for which this form is being completed: 🕮 Month | | Day | | | Year 2 | 0 | | |

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

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

Chediak-Higashi Syndrome Pre-HSCT Data

Registry Use Only

Sequence Number:

Date Received:

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 10.

1. What was the date of diagnosis of Chediak-Higashi Syndrome? Month | | Day | | | Year

2. Was genetic testing used to confirm the diagnosis?
   1 ☐ yes → Specify genetic mutation(s) identified:
   2 ☐ no
   3 ☐ unknown

   3. 1 ☐ yes 2 ☐ no CHS1 (LYST)
   4. 1 ☐ yes 2 ☐ no Other mutation → 5. Specify: _____

Pre-HSCT Data

Indicate which of the following manifestations of Chediak-Higashi Syndrome were present at any time prior to conditioning:

6. Leukocyte granules
   1 ☐ present
   2 ☐ absent
   3 ☐ unknown
7. Neutropenia (ANC < 1 x 10^9/L)
   1 ☐ present
   2 ☐ absent
   3 ☐ unknown
8. Oculocutaneous albinism
   1 ☐ present
   2 ☐ absent
   3 ☐ unknown
9. Recurrent infections
   1 ☐ present
   2 ☐ absent
   3 ☐ unknown

CIBMTR Form 2032 (CHS) v1.0 (1–4) July 2007

For internal use only: Document F00536 version 1.0 Replaces: n/a

Mail this form to your designated campus (Milwaukee or Minneapolis). Retain the original at the transplant center.

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10. Bleeding diathesis
- 1 ☐ present → Specify site:
  - 11. 1 ☐ yes 2 ☐ no Bleeding from the GI tract
  - 12. 1 ☐ yes 2 ☐ no Easy bruising
  - 13. 1 ☐ yes 2 ☐ no Hematuria
  - 14. 1 ☐ yes 2 ☐ no Oral bleeding
  - 15. 1 ☐ yes 2 ☐ no Recurrent nosebleeds
  - 16. 1 ☐ yes 2 ☐ no Other bleeding → 17. Specify site: _____
- 2 ☐ absent
- 3 ☐ unknown

18. Neurologic dysfuction
- 1 ☐ present → Specify site:
  - 19. 1 ☐ yes 2 ☐ no Abnormal gait
  - 20. 1 ☐ yes 2 ☐ no Developmental delay
  - 21. 1 ☐ yes 2 ☐ no Mental retardation
  - 22. 1 ☐ yes 2 ☐ no Motor weakness
  - 23. 1 ☐ yes 2 ☐ no Nystagmus
  - 24. 1 ☐ yes 2 ☐ no Seizures
  - 25. 1 ☐ yes 2 ☐ no Sensory deficits
  - 26. 1 ☐ yes 2 ☐ no Other dysfunction → 27. Specify site: _____
- 2 ☐ absent
- 3 ☐ unknown

Accelerated Phase

28. Did the recipient develop features of an accelerated phase at any time prior to the preparative regimen?
- 1 ☐ yes →
  - 29. Date accelerated phase was detected: Month | | Day | | Year | | | | ☐ date unknown
  - Specify accelerated feature(s) present:
    - 30. 1 ☐ present 2 ☐ absent 3 ☐ unknown Abnormal CSF (↑ WBC, ↑ protein)
    - 31. 1 ☐ present 2 ☐ absent 3 ☐ unknown Abnormal liver function
    - 32. 1 ☐ present 2 ☐ absent 3 ☐ unknown Anemia (Hb < 10 g/dL)
    - 33. 1 ☐ present 2 ☐ absent 3 ☐ unknown CMV associated with accelerated phase
    - 34. 1 ☐ present 2 ☐ absent 3 ☐ unknown EBV associated with accelerated phase
    - 35. 1 ☐ present 2 ☐ absent 3 ☐ unknown Fevers
    - 36. 1 ☐ present 2 ☐ absent 3 ☐ unknown Hemophagocytopenia (100 x 10^9/L)
    - 37. 1 ☐ present 2 ☐ absent 3 ☐ unknown Hepatomegaly
    - 38. 1 ☐ present 2 ☐ absent 3 ☐ unknown Increased triglycerides
    - 39. 1 ☐ present 2 ☐ absent 3 ☐ unknown Low fibrinogen
    - 40. 1 ☐ present 2 ☐ absent 3 ☐ unknown Lymphadenopathy
    - 41. 1 ☐ present 2 ☐ absent 3 ☐ unknown Neurologic dysfuction
    - 42. 1 ☐ present 2 ☐ absent 3 ☐ unknown Neutropenia (ANC < 1 x 10^9/L)
    - 43. 1 ☐ present 2 ☐ absent 3 ☐ unknown Splenomegaly
    - 44. 1 ☐ present 2 ☐ absent 3 ☐ unknown Thrombocytosis
    - 45. 1 ☐ present 2 ☐ absent 3 ☐ unknown Other infection associated with accelerated phase → 46. If yes, specify other infection: _____
- 2 ☐ no
- 3 ☐ unknown

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Specific Therapies Administered Prior to the Preparative Regimen

47. Did the recipient undergo treatment for accelerated phase prior to the preparative regimen?
    1 ☐ yes → Specify treatment(s) given:
    2 ☐ no
    3 ☐ unknown

48. 1 ☐ yes 2 ☐ no 3 ☐ unknown Acyclovir
49. 1 ☐ yes 2 ☐ no 3 ☐ unknown Antithymocyte globulin (ATG)
50. 1 ☐ yes 2 ☐ no 3 ☐ unknown Corticosteroids
51. 1 ☐ yes 2 ☐ no 3 ☐ unknown Etoposide (VP16)
52. 1 ☐ yes 2 ☐ no 3 ☐ unknown Ganciclovir (DHPG)
53. 1 ☐ yes 2 ☐ no 3 ☐ unknown Intrathecal methotrexate
54. 1 ☐ yes 2 ☐ no 3 ☐ unknown Intravenous immune globulin (IVIG)
55. 1 ☐ yes 2 ☐ no 3 ☐ unknown Interferon
56. 1 ☐ yes 2 ☐ no 3 ☐ unknown Other therapy →

57. Specify other therapy: _____

Clinical Status Immediately Prior to the Preparative Regimen

58. Did the recipient have magnetic resonance imaging (MRI) of the brain immediately prior to the preparative regimen?
    1 ☐ yes →
    2 ☐ no
    3 ☐ unknown

59. Specify MRI findings: _____

60. Is a copy of the MRI report attached?
    1 ☐ yes
    2 ☐ no

61. What was the disease status of Chediak-Higashi syndrome immediately prior to the preparative regimen?
    1 ☐ no prior accelerated phase
    2 ☐ in remission from accelerated phase
    3 ☐ in accelerated phase
    4 ☐ unknown

Most Recent Evaluation of Immunologic Function Prior to the Preparative Regimen

("Absent" is defined as ≤ 10% of normal value; "decreased" is defined as 11–50% of normal value.)

	Absent	Decreased	Normal	Increased	Not tested		Date of test Month	Day	Year	Date unknown
62. Cytotoxic T-cell activity	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	63.				☐
64. Granulocyte chemotaxis	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	65.				☐
66. IgG	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	67.				☐
68. IgA	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	69.				☐
70. IgM	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	71.				☐
72. IgE	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	73.				☐
74. Natural killer cell activity	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	75.				☐
76. T-cell function	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	77.				☐
78. T-cell numbers / subsets	1 ☐	2 ☐	3 ☐	4 ☐	5 ☐	79.				☐

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80. Were any other immunologic evaluations performed immediately prior to the preparative regimen?

1 ☐ yes → 81. Specify test and results: _____
2 ☐ no
3 ☐ unknown

82. Did the recipient receive IVIg infusions within 2 months prior to the above immunoglobulin measurement?

1 ☐ yes
2 ☐ no
3 ☐ unknown

83. Signed: _____
Person completing form

Please print name: _____

Phone: (_____) _____

Fax: (_____) _____

E-mail address: _____