



Chediak-Higashi Syndrome Pre-HSCT Data

Registry Use Only

Sequence
Number:

Date
Received:

CIBMTR Center Number:

CIBMTR Recipient ID:

Today's Date: / / (2 0)

Date of HSCT for which this form is being completed: / / (2 0)

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 10.

1. What was the date of diagnosis of Chediak-Higashi Syndrome? / /
2. Was genetic testing used to confirm the diagnosis?

- 1 yes
2 no
3 unknown

Specify genetic mutation(s) identified:

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 10⁹/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

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
- 2 absent
- 3 unknown

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:

18. Neurologic dysfunction

- 1 present
- 2 absent
- 3 unknown

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:

Accelerated Phase

28. Did the recipient develop features of an accelerated phase at any time prior to the preparative regimen?

- 1 yes
- 2 no
- 3 unknown

29. Date accelerated phase was detected: date unknown
Month Day Year

Specify accelerated feature(s) present:

- 30. 1 present 2 absent 3 unknown Abnormal CSF (\uparrow WBC, \uparrow 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 ($100 \times 10^9/L$)
- 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 dysfunction
- 42. 1 present 2 absent 3 unknown Neutropenia ($ANC < 1 \times 10^9/L$)
- 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:

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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
- 2 no
- 3 unknown

Specify treatment(s) given:

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		Month	Date of test		Year	Date unknown
							Day				
62. Cytotoxic T-cell activity	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	63.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
64. Granulocyte chemotaxis	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	65.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
66. IgG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	67.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
68. IgA	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	69.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
70. IgM	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	71.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
72. IgE	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	73.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
74. Natural killer cell activity	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	75.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
76. T-cell function	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	77.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
78. T-cell numbers / subsets	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	79.	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>

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

- 1 yes
- 2 no
- 3 unknown

81. Specify test and results:

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: _____