

**INSERT XIX**  
**Chediak-Higashi Syndrome**

TEAM

IUBMID

(Institutional Unique Blood or Marrow Transplant Identification Number)

Date of transplant for which this form is being completed:     
Month Day Year

**FOR REGISTRY USE ONLY:**

I.D. --

Date received:

Registry: IBMTR ABMTR (circle one)

Date of report:     
Month Day Year

**Epidemiologic Data**

\* If this is a report of a second (or subsequent) transplant, check here  and go to Q.59

1. Is there a family history of Chediak-Higashi Syndrome?

- 1  Yes  
0  No  
8  Unknown

Specify affected members:

- | <u>Yes</u>                 | <u>No</u>                  |                       |
|----------------------------|----------------------------|-----------------------|
| 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Sibling(s)            |
| 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Cousin(s)             |
| 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Aunt(s)/Uncle(s)      |
| 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Other, specify: _____ |

2. Is there a history of consanguinity?

- 1  Yes  
0  No  
8  Unknown

**Pretransplant Information**

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

- |   |                                    |                                   |                                    |
|---|------------------------------------|-----------------------------------|------------------------------------|
| 3. Leukocyte granules                     | 1 <input type="checkbox"/> Present | 0 <input type="checkbox"/> Absent | 8 <input type="checkbox"/> Unknown |
| 4. Neutropenia (ANC<1x10 <sup>9</sup> /L) | 1 <input type="checkbox"/> Present | 0 <input type="checkbox"/> Absent | 8 <input type="checkbox"/> Unknown |
| 5. Oculocutaneous albinism                | 1 <input type="checkbox"/> Present | 0 <input type="checkbox"/> Absent | 8 <input type="checkbox"/> Unknown |
| 6. Recurrent infections                   | 1 <input type="checkbox"/> Present | 0 <input type="checkbox"/> Absent | 8 <input type="checkbox"/> Unknown |
| 7. Bleeding diatheses                     | 1 <input type="checkbox"/> Present | 0 <input type="checkbox"/> Absent | 8 <input type="checkbox"/> Unknown |

Specify:

- | <u>Yes</u>                     | <u>No</u>                  |                      | <u>Yes</u>                     | <u>No</u>                  |                            |
|--------------------------------|----------------------------|----------------------|--------------------------------|----------------------------|----------------------------|
| 8. 1 <input type="checkbox"/>  | 0 <input type="checkbox"/> | Easy bruising        | 11. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Bleeding from the GI tract |
| 9. 1 <input type="checkbox"/>  | 0 <input type="checkbox"/> | Recurrent nosebleeds | 12. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Hematuria                  |
| 10. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Oral bleeding        | 13. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Other, specify: _____      |

14. Neurologic dysfunction 1  Present 0  Absent 8  Unknown

Specify:

- | <u>Yes</u>                     | <u>No</u>                  |                     | <u>Yes</u>                     | <u>No</u>                  |                       |
|--------------------------------|----------------------------|---------------------|--------------------------------|----------------------------|-----------------------|
| 15. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Seizures            | 19. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Abnormal gait         |
| 16. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Mental retardation  | 20. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Motor weakness        |
| 17. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Developmental delay | 21. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Sensory deficits      |
| 18. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Nystagmus           | 22. 1 <input type="checkbox"/> | 0 <input type="checkbox"/> | Other, specify: _____ |

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**Accelerated phase:**

23. Did patient develop features of an accelerated phase at any time prior to conditioning?

- 1  Yes
- 0  No
- 8  Unknown

Specify:

24. Date:        Date unknown  
Month Day Year

Indicate whether each of the following were present:

	<u>Present</u>	<u>Absent</u>	<u>Unknown</u>
25. Fevers	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
26. Splenomegaly	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
27. Hepatomegaly	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
28. Lymphadenopathy	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
29. Neurologic dysfunction	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
30. Abnormal liver function	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
31. Abnormal CSF (↑ WBC, ↑ protein)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
32. Anemia (Hb<10g/dl)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
33. Neutropenia (ANC<1x10 <sup>9</sup> /L)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
34. Thrombocytopenia (<100x10 <sup>9</sup> /L)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
35. Hemophagocytosis	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
36. Increased triglycerides	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
37. Low fibrinogen	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
38. EBV associated with accelerated phase	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
39. CMV associated with accelerated phase	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
40. Other infection associated with accelerated phase (specify: _____)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>

**Specific therapies administered prior to conditioning:**

- |  | <u>Unknown</u>             | <u>No</u>                  | <u>Yes</u>                 |  |
|--|----------------------------|----------------------------|----------------------------|--|
| 41. Corticosteroids                    | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 42. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 43. Intravenous immune globulin (IVIg) | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 44. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 45. Intrathecal methotrexate           | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 46. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 47. Antithymocyte globulin (ATG)       | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 48. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 49. Etoposide (VP16)                   | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 50. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 51. Interferon                         | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 52. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 53. Acyclovir                          | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 54. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 55. Ganciclovir (DHPG)                 | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 56. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| 57. Other, specify: _____              | 8 <input type="checkbox"/> | 0 <input type="checkbox"/> | 1 <input type="checkbox"/> | 58. Did patient respond? 1 <input type="checkbox"/> Yes 0 <input type="checkbox"/> No 8 <input type="checkbox"/> Unknown |
| _____                                  |                            |                            |                            |  |
| _____                                  |                            |                            |                            |  |
| _____                                  |                            |                            |                            |  |

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**Clinical status immediately prior to conditioning:**

- Specify Units**                      **Unknown**
59. Hemoglobin:           1  g/dL 2  g/L 3  mmol/L
60. WBC:           1  x 10<sup>9</sup>/L 2  x 10<sup>6</sup>/L
61. Platelets:           1  x 10<sup>9</sup>/L 2  x 10<sup>6</sup>/L
62. Absolute neutrophil count:           1  x 10<sup>9</sup>/L 2  x 10<sup>6</sup>/L
63. Hepatomegaly 1  Yes 0  No
64. Splenomegaly 1  Yes 0  No

65. Did the patient develop a malignancy at any time prior to conditioning?  
 1  Yes  Specify histology and site: \_\_\_\_\_  
 0  No  
 8  Unknown

66. Did the patient have magnetic resonance imaging (MRI) of brain prior to conditioning?  
 1  Yes  Specify findings: \_\_\_\_\_  
 0  No  
 8  Unknown

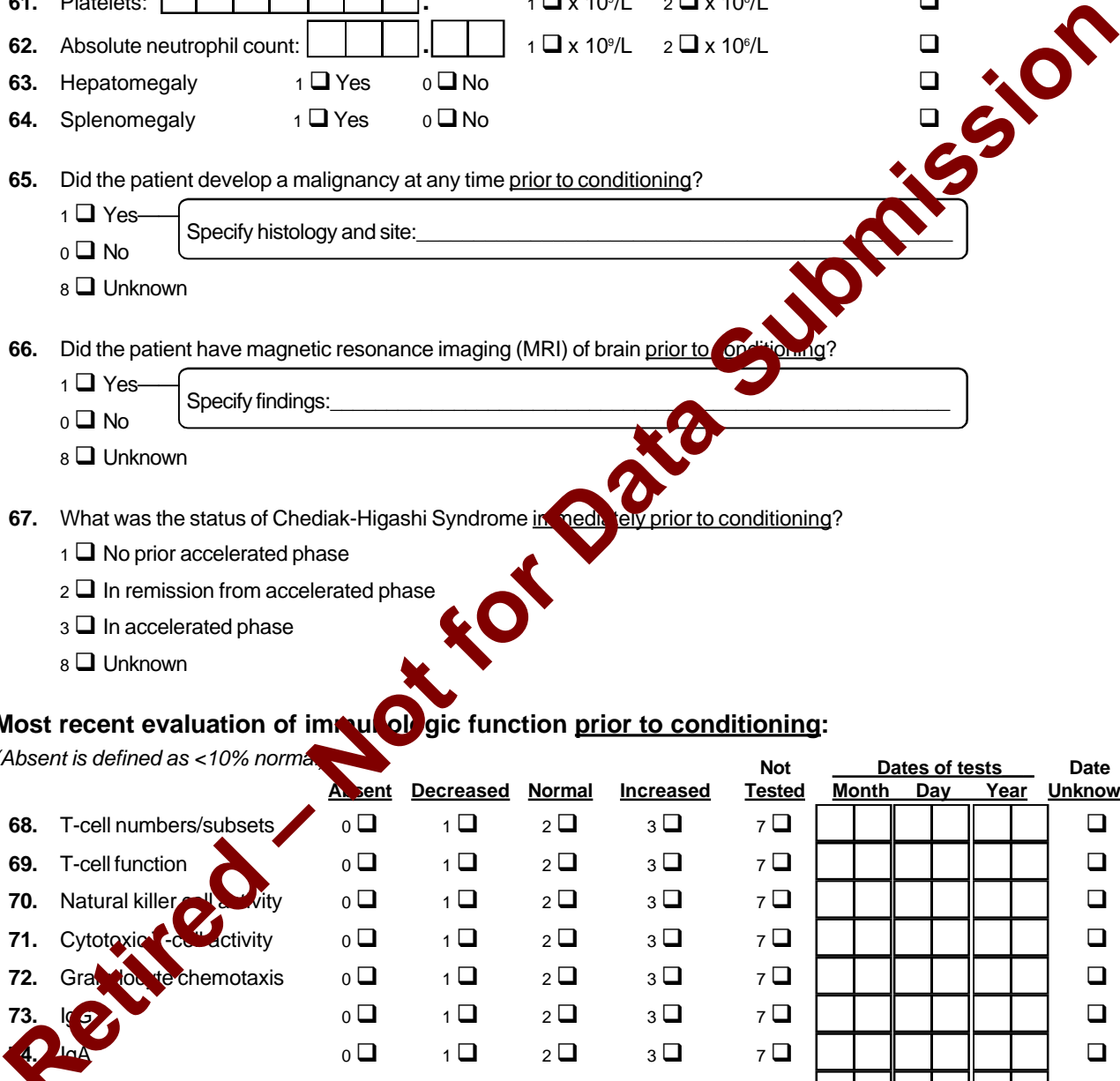
67. What was the status of Chediak-Higashi Syndrome immediately prior to conditioning?  
 1  No prior accelerated phase  
 2  In remission from accelerated phase  
 3  In accelerated phase  
 8  Unknown

**Most recent evaluation of immunologic function prior to conditioning:**

(Absent is defined as <10% normal)

	Absent	Decreased	Normal	Increased	Not Tested	Dates of tests			Date Unknown
						Month	Day	Year	
68. T-cell numbers/subsets	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
69. T-cell function	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
70. Natural killer cell activity	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
71. Cytotoxic T-cell activity	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
72. Granulocyte chemotaxis	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
73. IgG	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
74. IgA	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
75. IgM	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
76. IgE	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>

77. Other immunologic evaluation?  
 1  Yes  Specify tests and results: \_\_\_\_\_  
 0  No



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### Posttransplant Information

**To be completed 100 days posttransplant, or at time of death if death occurred <100 days posttransplant, or immediately prior to start of high-dose therapy (conditioning) for second transplant if second transplant done <100 days after first transplant.**

#### Posttransplant follow-up:

78. Current status of clinical features of Chediak-Higashi Syndrome (status at 100 day visit or at time of death if died <100 days posttransplant):

- 0  Absent
- 3  Persistent
- 4  Recurrent

Specify:

79. Date of recurrence:     Date unknown  
Month Day Year

	Present	Absent	Unknown
80. Leukocyte granules	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
81. Splenomegaly	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
82. Hepatomegaly	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
83. Anemia (Hb<10g/dl)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
84. Neutropenia (ANC<1x10 <sup>9</sup> /L)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
85. Thrombocytopenia (<100x10 <sup>9</sup> /L)	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
86. Recurrent infections	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>
87. Bleeding diathesis	1 <input type="checkbox"/>	0 <input type="checkbox"/>	8 <input type="checkbox"/>

88. Neurologic symptoms of Chediak-Higashi Syndrome posttransplant:

- 0  Absent
- 1  Present
- 8  Unknown

89. Neurologic symptoms were present pretransplant?  
 1  Yes 0  No (symptoms are newly developed) 8  Unknown

90. Current status of neurologic dysfunction compared to pretransplant:  
 1  Improved 2  Unchanged 3  Worse 8  Unknown

91. Accelerated phase posttransplant?

- 1  Yes
- 0  No

92. Date:     
Month Day Year

93. Date current status evaluated:

Month Day Year

#### Current assessment of immunologic function posttransplant (assessment at 100 day visit or at time of death if died <100 days posttransplant):

(Absent is defined as <10% normal)

	Absent	Decreased	Normal	Increased	Not Tested	Dates of tests			Date Unknown
						Month	Day	Year	
94. T-cell numbers subsets	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
95. T-cell function	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
96. Natural killer cell activity	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
97. Cytotoxic T-cell activity	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
98. Granulocyte chemotaxis	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
99. IgG	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
100. IgA	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
101. IgM	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>
102. IgE	0 <input type="checkbox"/>	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	7 <input type="checkbox"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="checkbox"/>

103. Was the patient receiving IVIG at the time immunoglobulin levels were measured?

- 1  Yes
- 0  No
- 8  Unknown