

National Marrow Donor Program®  
**Insert XV – Chediak-Higashi Syndrome**

**Registry Use Only**

Sequence Number:

Date Received:

**Unrelated**

Recipient NMDP ID:  -  -

Recipient Last Name:

Recipient Local ID (optional):

Today's Date:  /  /  TC Code:

Month                  Day                  Year

Date of Transplant for which this form is being completed:  /  /

Month                  Day                  Year

Product type:  Marrow (Form 120)     PBSC (Form 520)     Cord blood (Form 620)

**This form must be accompanied by Form 120, 520, 620 – Recipient Baseline and Transplant Data. All information in the box above, including the date, should be identical with the corresponding Form 120, 520, 620. Information should come from an actual examination by the Transplant Center physician, or abstraction of the recipient's medical records.**

**Epidemiologic Data**

**If this is a report of a second (or subsequent) transplant, check box 1 and go to question 33.**

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

- 1  yes
- 2  no
- 3  unknown

2. Specify affected member(s).

- a. Sibling(s)                  1  yes    2  no
- b. Cousins                  1  yes    2  no
- c. Aunt(s), Uncle(s)        1  yes    2  no
- d. Other                      1  yes    2  no

If yes, specify: \_\_\_\_\_

3. Is there a history of consanguinity?

- 1  yes
- 2  no
- 3  unknown

**Pre-Transplant Data**

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

- 4. Leukocyte granules
  - 1  present
  - 2  absent
  - 3  unknown
- 5. Neutropenia (ANC < 1 x 10<sup>9</sup>/L)
  - 1  present
  - 2  absent
  - 3  unknown
- 6. Oculocutaneous albinism
  - 1  present
  - 2  absent
  - 3  unknown
- 7. Recurrent infections
  - 1  present
  - 2  absent
  - 3  unknown

**Mail to NMDP Registry with Form 120, 520, 620. Retain a copy at the transplant center.**

Recipient NMDP ID:    -    -

Recipient Last Name:

8. Bleeding diathesis

- 1  present
- 2  absent
- 3  unknown

9. Specify:

- a. Easy bruising 1  yes 2  no
- b. Recurrent nosebleeds 1  yes 2  no
- c. Oral bleeding 1  yes 2  no
- d. Bleeding from the GI tract 1  yes 2  no
- e. Hematuria 1  yes 2  no
- f. Other, specify: \_\_\_\_\_ 1  yes 2  no

10. Neurologic dysfunction

- 1  present
- 2  absent
- 3  unknown

11. Specify:

- a. Seizures 1  yes 2  no
- b. Mental retardation 1  yes 2  no
- c. Developmental delay 1  yes 2  no
- d. Nystagmus 1  yes 2  no
- e. Abnormal gait 1  yes 2  no
- f. Motor weakness 1  yes 2  no
- g. Sensory deficits 1  yes 2  no
- f. Other, specify: \_\_\_\_\_ 1  yes 2  no

**Accelerated Phase**

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

- 1  yes
- 2  no
- 3  unknown

13. Date:   /   /      date unknown  
Month Day Year

14. Indicate whether each of the following were present:

- a. Fevers 1  present 2  absent 3  unknown
- b. Splenomegaly 1  present 2  absent 3  unknown
- c. Hepatomegaly 1  present 2  absent 3  unknown
- d. Lymphadenopathy 1  present 2  absent 3  unknown
- e. Neurologic dysfunction 1  present 2  absent 3  unknown
- f. Abnormal liver function 1  present 2  absent 3  unknown
- g. Abnormal CSF ( $\uparrow$  WBC,  $\uparrow$  protein) 1  present 2  absent 3  unknown
- h. Anemia (Hb < 10 g/dL) 1  present 2  absent 3  unknown
- i. Neutropenia (ANC <  $1 \times 10^9/L$ ) 1  present 2  absent 3  unknown
- j. Thrombocytosis 1  present 2  absent 3  unknown
- k. Hemophagocytopenia ( $100 \times 10^9/L$ ) 1  present 2  absent 3  unknown
- l. Increased triglycerides 1  present 2  absent 3  unknown
- m. Low fibrinogen 1  present 2  absent 3  unknown
- n. EBV associated with accelerated phase 1  present 2  absent 3  unknown
- o. CMV associated with accelerated phase 1  present 2  absent 3  unknown
- p. Other infection associated with accelerated phase (Specify: \_\_\_\_\_) 1  present 2  absent 3  unknown

Recipient NMDP ID:    -    -

Recipient Last Name:

### Specific Therapies Administered Prior to Conditioning

15. Corticosteroids

- 1  yes  
2  no  
3  unknown

16. Did patient respond?

- 1  yes 2  no 3  unknown

17. Intravenous immune globulin (IVIG)

- 1  yes  
2  no  
3  unknown

18. Did patient respond?

- 1  yes 2  no 3  unknown

19. Intrathecal methotrexate

- 1  yes  
2  no  
3  unknown

20. Did patient respond?

- 1  yes 2  no 3  unknown

21. Antithymocyte globulin (ATG)

- 1  yes  
2  no  
3  unknown

22. Did patient respond?

- 1  yes 2  no 3  unknown

23. Etoposide (VP16)

- 1  yes  
2  no  
3  unknown

24. Did patient respond?

- 1  yes 2  no 3  unknown

25. Interferon

- 1  yes  
2  no  
3  unknown

26. Did patient respond?

- 1  yes 2  no 3  unknown

27. Acyclovir

- 1  yes  
2  no  
3  unknown

28. Did patient respond?

- 1  yes 2  no 3  unknown

29. Ganciclovir (DHPG)

- 1  yes  
2  no  
3  unknown

30. Did patient respond?

- 1  yes 2  no 3  unknown

31. Other, specify drug: \_\_\_\_\_

- 1  yes  
2  no  
3  unknown

32. Did patient respond?

- 1  yes 2  no 3  unknown

Recipient NMDP ID: --

Recipient Last Name:

### Clinical Status Immediately Prior to Conditioning

33. Hemoglobin:  •  1  g/dL 2  g/L 3  mmol/L 4  unknown

34. WBC:  •  x 10<sup>9</sup>/L  unknown

35. Platelets:  •  x 10<sup>9</sup>/L  unknown

36. Absolute neutrophil count:  •  x 10<sup>9</sup>/L  unknown

37. Hepatomegaly:  
1  yes  
2  no  
3  unknown

38. Splenomegaly:  
1  yes  
2  no  
3  unknown

39. Did the patient develop a malignancy at any time prior to conditioning?

- 1  yes
- 2  no
- 3  unknown

40. Specify histology and site:

41. Did the patient have magnetic resonance imaging (MRI) of brain prior to conditioning?

- 1  yes
- 2  no
- 3  unknown

42. Specify findings:

43. 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
- 3  unknown

Recipient NMDP ID:    -    -

Recipient Last Name:

### Most Recent Evaluation of Immunologic Function Prior to Conditioning

(Absent is defined as <10% normal)

	Absent	Decreased	Normal	Increased	Not tested	Date of test			Date unknown
44. T-cell numbers/subsets	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
45. T-cell function	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
46. Natural killer cell activity	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
47. Cytotoxic T-cell activity	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
48. Granulocyte chemotaxis	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
49. IgG	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
50. IgA	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
51. IgM	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>
52. IgE	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	5 <input type="checkbox"/>	<input type="text"/> Month	<input type="text"/> Day	<input type="text"/> <input type="text"/> <input type="text"/> Year	<input type="checkbox"/>

53. Other immunologic evaluation?

- 1  yes  
2  no

54. Specify tests and results: \_\_\_\_\_  
 \_\_\_\_\_  
 \_\_\_\_\_

**Continue with question 10 on page 5 of Form 120, 520, 620.**