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

3. 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 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
### 10. Bleeding diathesis

- **Specify site:**
  - 11. yes 2 no Bleeding from the GI tract
  - 12. yes 2 no Easy bruising
  - 13. yes 2 no Hematuria
  - 14. yes 2 no Oral bleeding
  - 15. yes 2 no Recurrent nosebleeds
  - 16. yes 2 no Other bleeding

### 18. Neurologic dysfunction

- **Specify site:**
  - 19. yes 2 no Abnormal gait
  - 20. yes 2 no Developmental delay
  - 21. yes 2 no Mental retardation
  - 22. yes 2 no Motor weakness
  - 23. yes 2 no Nystagmus
  - 24. yes 2 no Seizures
  - 25. yes 2 no Sensory deficits
  - 26. yes 2 no Other dysfunction

### Accelerated Phase

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

- 1 yes 2 no

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

- **Specify accelerated feature(s) present:**
  - 30. present 2 absent 3 unknown Abnormal CSF (↑ WBC, ↑ protein)
  - 31. present 2 absent 3 unknown Abnormal liver function
  - 32. present 2 absent 3 unknown Anemia (Hb < 10 g/dL)
  - 33. present 2 absent 3 unknown CMV associated with accelerated phase
  - 34. present 2 absent 3 unknown EBV associated with accelerated phase
  - 35. present 2 absent 3 unknown Fevers
  - 36. present 2 absent 3 unknown Hemophagocytopenia (100 x 10⁹/L)
  - 37. present 2 absent 3 unknown Hepatomegaly
  - 38. present 2 absent 3 unknown Increased triglycerides
  - 39. present 2 absent 3 unknown Low fibrinogen
  - 40. present 2 absent 3 unknown Lymphadenopathy
  - 41. present 2 absent 3 unknown Neurologic dysfunction
  - 42. present 2 absent 3 unknown Neutropenia (ANC < 1 x 10⁹/L)
  - 43. present 2 absent 3 unknown Splenomegaly
  - 44. present 2 absent 3 unknown Thrombocytosis
  - 45. present 2 absent 3 unknown Other infection associated with accelerated phase

46. If yes, specify other infection:  
   -
### 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. [ ] yes 2. [ ] no 3. [ ] unknown Acyclovir
- 49. [ ] yes 2. [ ] no 3. [ ] unknown Antithymocyte globulin (ATG)
- 50. [ ] yes 2. [ ] no 3. [ ] unknown Corticosteroids
- 51. [ ] yes 2. [ ] no 3. [ ] unknown Etoposide (VP16)
- 52. [ ] yes 2. [ ] no 3. [ ] unknown Ganciclovir (DHPG)
- 53. [ ] yes 2. [ ] no 3. [ ] unknown Intrathecal methotrexate
- 54. [ ] yes 2. [ ] no 3. [ ] unknown Intravenous immune globulin (IVIG)
- 55. [ ] yes 2. [ ] no 3. [ ] unknown Interferon
- 56. [ ] yes 2. [ ] no 3. [ ] unknown 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

### 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.)

<table>
<thead>
<tr>
<th>Test</th>
<th>Absent</th>
<th>Decreased</th>
<th>Normal</th>
<th>Increased</th>
<th>Not tested</th>
<th>Month</th>
<th>Date of test</th>
<th>Year</th>
<th>Date unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>62. Cytotoxic T-cell activity</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>63. [ ]</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>64. Granulocyte chemotaxis</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>65. [ ]</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>66. IgG</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>67. [ ]</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>68. IgA</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>69. [ ]</td>
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</tr>
<tr>
<td>70. IgM</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>71. [ ]</td>
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<tr>
<td>72. IgE</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>73. [ ]</td>
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</tr>
<tr>
<td>74. Natural killer cell activity</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>75. [ ]</td>
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<tr>
<td>76. T-cell function</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>77. [ ]</td>
<td></td>
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</tr>
<tr>
<td>78. T-cell numbers / subsets</td>
<td>1 [ ]</td>
<td>2 [ ]</td>
<td>3 [ ]</td>
<td>4 [ ]</td>
<td>5 [ ]</td>
<td>79. [ ]</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
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: ______________________________________________