1. What was the date of diagnosis of Hemophagocytic Lymphohistiocytosis?  
2. Is there a family history of hemophagocytic disorders?  
3. 1 yes 2 no Aunt(s) / uncle(s)  
4. 1 yes 2 no Cousin(s)  
5. 1 yes 2 no Sibling(s)  
6. 1 yes 2 no Other  
7. Specify relationship:  
8. Is there a family history of consanguinity (descent from common ancestors / inter-familial marriage)?  
9. Describe:  
10. Specify the following clinical and laboratory features at diagnosis:  
   - Anemia (hemoglobin < 10 g/dL)  
   - Fever (> 38.5°C for > 7 days within 1 week of diagnosis)  
   - Hepatomegaly (> 3 cm below right costal margin)  
   - Hypertriglyceridemia (> 200 mg/dL)  
   - Hypofibrinogenemia (< 150 mg/dL)  
   - Neutropenia (ANC < 1.0 x 10⁹/L)  
   - Splenomegaly (> 3 cm below left costal margin)  
   - Thrombocytopenia (< 100 x 10⁹/L)  

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 90.

Specify affected member(s):  
3. 1 yes 2 no Aun(t(s) / uncle(s)  
4. 1 yes 2 no Cousin(s)  
5. 1 yes 2 no Sibling(s)  
6. 1 yes 2 no Other  

Mail this form to your designated campus (Milwaukee or Minneapolis). Retain the original at the transplant center.
Specify the cerebrospinal fluid findings at diagnosis:
18. Neopterin level  1  normal  2  elevated  3  not tested
19. Protein  1  normal  2  elevated  3  not tested
20. WBC count  1  ≤ 5 cells/µl  2  > 5 cells/µl  3  not tested

21. Was there evidence of hemophagocytosis in the cerebrospinal fluid at diagnosis?
   1  yes  2  no  3  not tested

22. Were central nervous system (CNS) abnormalities found on computed tomography (CT or CAT) or magnetic resonance imaging (MRI) scans at any time prior to the preparative regimen?
   1  yes  2  no  3  unknown / not tested

Specify type of scan performed:
23. 1  yes  2  no CT
24. Specify abnormality detected on CT: __________________
25. 1  yes  2  no MRI
26. Specify abnormality detected on MRI: __________________
27. Is a copy of the report(s) attached?
   1  yes  2  no

28. Were there any clinical neurologic abnormalities at any time prior to the preparative regimen?
   1  yes  2  no  3  unknown

Specify abnormalities:
29. 1  yes  2  no Abnormal gait
30. 1  yes  2  no Developmental delay
31. 1  yes  2  no Mental retardation
32. 1  yes  2  no Motor weakness
33. 1  yes  2  no Seizures
34. 1  yes  2  no Sensory deficits
35. 1  yes  2  no Other
36. Specify abnormality: __________________

Infection History at Time of Presentation with Disease

37. Was an infection documented at diagnosis?
   1  yes  2  no

Specify infection(s):
38. Cytomegalovirus (CMV):
   1  yes  2  no
39. Specify the test method used for diagnosis of CMV:
   1  culture
   2  polymerase chain reaction (PCR)
40. Epstein-Barr virus (EBV):
   1  yes  2  no
41. Specify the test method used for diagnosis of EBV:
   1  in situ hybridization
   2  PCR
   3  serology
   Specify titers:
   42. EBNA: [ ] [ ] [ ] [ ] [ ]
   43. Early antigen: [ ] [ ] [ ] [ ] [ ]
   44. Viral capsid IgG: [ ] [ ] [ ] [ ] [ ]
   45. Viral capsid IgM: [ ] [ ] [ ] [ ] [ ]
ERROR CORRECTION FORM

Sequence Number: 

Today’s Date: 

Infusion Date: 

CIBMTR Recipient ID: 

CIBMTR Center Number: 

Infusion Date: 

Month Day Year 

CIBMTR Center Number: 

CIBMTR Recipient ID: 

Initials: 

Today's Date: 

Month Day Year 

Infusion Date: 

Month Day Year 

Codes for Commonly Reported Organisms

Bacterial Infections
121 Acinetobacter
122 Actinomycetes
123 Bacillus
124 Bacteroides (fragilis, uniformis, vulgaris, other species)
125 Bordetella pertussis (whooping cough)
126 Borrelia (Lyme disease)
127 Brachymania or Moraxella catarrhalis (other species)
128 Campylobacter (all species)
129 Capnocytophaga
171 Chlamydia pneumoniae
172 Other chlamydia, specify
173 Chlamydia, NOS
130 Clostridium difficile
131 Clostridium (all species except difficile)
132 Clostridium difficile
133 Corynebacterium jeikeium
134 Corynebacterium (all non-diptheria species)
101 Coxiella
135 Enterobacter
177 Enterococcus, vancomycin resistant (VRE)
136 Escherichia (also E. coli)
137 Flavimona argyribanites
138 Flavobacterium
139 Fusobacterium
144 Haemophilus (all species, including influenzae)
145 Helicobacter pylori
146 Klebsiella
147 Lactobacillus (bulgaricus, acidophilus, other species)
102 Legionella
103 Lepotiricia buccalis
148 Leptotrichia buccalis
149 Leuconostoc (all species)
104 Listeria
150 Lactobacillus (bulgaricus, acidophilus, other species)
151 Micrococcus, NOS
152 Neisseria (gonorrhoea, meningitidis, other species)
153 Pasteurella multocida
154 Propionibacterium (acnes, avidum, granulomus, other species)
155 Proteus
156 Pseudomonas (all species except cepacia & maltophilia)
157 Pseudomonas or Burkholderia cepacia
158 Pseudomonas or Xanthomonas maltophilia
159 Rhodococcus
160 Salmonella (all species)
161 Serratia marcescens
162 Shigella
163 Staphylococcus, coagulase negative (not aureus)
164 Staphylococcus aureus
165 Staphylococcus, NOS
166 Stomatococcus mucogalis
167 Streptococcus (all species except Enterococcus)
168 Streptococcus pneumoniae
169 Vibrio (all species)
197 Multiple bacteria at a single site, specify bacterial codes
198 Other bacteria, specify
199 Other bacteria, specify (see codes below)
501 Suspected atypical bacterial infection
502 Suspected bacterial infection
47. Specify other infection: (see codes below)
48. If other organism (code 198, 209, 259, 329, or 409), specify:
49. 1 yes 2 no Blood
50. 1 yes 2 no Cerebrospinal fluid (CSF)
51. 1 yes 2 no Urine
52. 1 yes 2 no Tissue biopsy
53. Specify tissue / site:
54. 1 yes 2 no Other body fluid
55. Specify fluid / site:

Fungal Infections
200 Candida, NOS
201 Candida albicans
202 Candida guillermondi
203 Candida parapsilosis
204 Candida tropicalis
205 Candida (Torulopsis) glabrata
206 Candida krusei
207 Candida lusitaniae
208 Candida lusitaniae
209 Other Candida, specify
210 Aspergillus, NOS
211 Aspergillus flavus
212 Aspergillus fumigatus
213 Aspergillus niger
214 Other Aspergillus, specify
220 Cryptococcus species
230 Fusarium species
260 Histoplasmosis
261 Histioplasmosis
262 Polymyces, NOS
263 Mucomycosis
242 Rhizopus
250 Yeast, NOS
298 Other fungus, specify
260 Pneumocystis (PCP / PJP)
503 Suspected fungal infection
504 Suspected fungal infection
505 Suspected viral infection

Viral Infections
301 Herpes simplex (HSV1, HSV2)
302 Varicella (herpes zoster, chicken pox)
303 Cytomegalovirus (CMV)
304 Adenovirus
305 Enterovirus (coxackie, echo, polio)
306 Hepatitis A (HAV)
307 Hepatitis B (HBV, Australian antigen)
308 Hepatitis C (HCV)
309 HIV-1 (HIV-III)
310 Influenza, NOS
311 Influenza A
312 Influenza B
313 Measles (rubeola)
314 Mumps
315 Progressive multifocal leukoencephalopathy (PML)
316 Respiratory syncytial virus (RSV)
317 Rubella (German measles)
318 Rubeola (German measles)
319 Human herpesvirus-6 (HHV-6)
320 Epstein-Barr virus (EBV)
321 Polyoma virus (BK virus, JC virus)
322 Human papilloma virus (HPV)
323 Other virus, specify
324 Other virus, specify
325 Other virus, specify
326 Other virus, specify
327 Other virus, specify
328 Other virus, specify
329 Other virus, specify
504 Suspected viral infection

Parasitic Infections
402 Toxoplasma
403 Giardia
404 Cryptosporidium
405 Other parasite, specify
505 Suspected parasite infection

Other Infections
509 No organism identified

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
Specify the site(s) where hemophagocytosis was documented at diagnosis:

<table>
<thead>
<tr>
<th>Present</th>
<th>Absent</th>
<th>Not tested</th>
<th>Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>56.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>57.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>58.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>59.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>60.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>61.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

62. Specify site of hemophagocytosis: ____________________________

63. What is the current natural killer cell function? (Refers to specific cytolysis of NK-sensitive target cells, e.g. K562.)
   1 □ absent (≤ 10% normal response)
   2 □ decreased (11–60% normal response)
   3 □ normal
   4 □ increased
   5 □ not tested

64. Was treatment given at any time prior to the preparative regimen?

<table>
<thead>
<tr>
<th>Present</th>
<th>Absent</th>
<th>Not tested</th>
<th>Therapy and reason for therapy:</th>
</tr>
</thead>
<tbody>
<tr>
<td>65.</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

65. Cyclosporine: Given for induction / maintenance? Given for disease relapse?
   1 □ yes  66. 1 □ yes 2 □ no
   2 □ no

66. Intrathecal methotrexate:
   1 □ yes  67. 1 □ yes 2 □ no
   2 □ no

71. IVIG:
   1 □ yes  72. 1 □ yes 2 □ no
   2 □ no

74. Steroids:
   1 □ yes  73. 1 □ yes 2 □ no
   2 □ no

84. Radiation therapy:
   1 □ yes  85. 1 □ yes 2 □ no
   2 □ no

87. Specify radiation field:

88. Specify total dose: ________ cGy

89. Was this therapy given following the HLH-94 protocol of the Histiocyte Society?
   1 □ yes
   2 □ no
   3 □ unknown
90. Was CNS disease quiescent (inactive) at any time prior to the preparative regimen?

1. yes
2. no
3. unknown

Specify:
91. yes 2 no 3 unknown Normal or stable CT or MRI of CNS
92. yes 2 no 3 unknown Normal CSF neopterin level
93. yes 2 no 3 unknown Normal CSF protein
94. yes 2 no 3 unknown Normal CSF WBC (< 5 cells/mm³)

95. Is a copy of the report attached?
1. yes
2. no

96. Was systemic disease quiescent at any time prior to the preparative regimen?

1. yes
2. no
3. unknown

Specify:
97. yes 2 no 3 unknown ANC > 1.0 x 10⁹/L (without growth factor support)
98. yes 2 no 3 unknown Hemoglobin > 9 g/dL without transfusion
99. yes 2 no 3 unknown Hepatomegaly resolved (≤ 3 cm below costal margin)
100. yes 2 no 3 unknown Normal fibrinogen
101. yes 2 no 3 unknown Normal triglycerides
102. yes 2 no 3 unknown Platelets > 100 x 10⁹/L without transfusion
103. yes 2 no 3 unknown Splenomegaly resolved (≤ 3 cm below costal margin)

104. Were there any signs of disease relapse / reactivation prior to HSCT?

1. yes
2. no

105. Specify the date of the first relapse / reactivation:

Month Day Year

106. Specify the site of the first relapse / reactivation:
1. CNS
2. systemic
3. both

107. Specify the date of the second relapse / reactivation:

Month Day Year

108. Specify the site of the second relapse / reactivation:
1. CNS
2. systemic
3. both

109. Specify the date of the third relapse / reactivation:

Month Day Year

110. Specify the site of the third relapse / reactivation:
1. CNS
2. systemic
3. both

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
Specify the clinical and laboratory features just prior to the preparative regimen:

111. 1 □ yes 2 □ no 3 □ unknown Anemia (hemoglobin < 10 g/dL)
112. 1 □ yes 2 □ no 3 □ unknown Fever (> 38.5°C for > 7 days within 1 week of conditioning)
113. 1 □ yes 2 □ no 3 □ unknown Hepatomegaly (> 3 cm below right costal margin)
114. 1 □ yes 2 □ no 3 □ unknown Hypertriglyceridemia (> 200 mg/dL)
115. 1 □ yes 2 □ no 3 □ unknown Hypofibrinogenemia (< 150 mg/dL)
116. 1 □ yes 2 □ no 3 □ unknown Neutropenia (ANC < 1.0 x 10^9/L)
117. 1 □ yes 2 □ no 3 □ unknown Splenomegaly (> 3 cm below left costal margin)
118. 1 □ yes 2 □ no 3 □ unknown Thrombocytopenia (< 100 x 10^9/L)

119. What was the status of CNS disease just prior to the preparative regimen?
   1 □ active
   2 □ non-active, quiescent
   3 □ CNS disease absent at diagnosis

120. Signed: __________________________

   Person completing form

   Please print name: __________________________

   Phone: (__________) __________________________

   Fax: (__________) __________________________

   E-mail address: __________________________