



# Hemophagocytic Lymphohistiocytosis 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  multiple cord blood units infused  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 90.

1. What was the date of diagnosis of Hemophagocytic Lymphohistiocytosis?  /  /

2. Is there a family history of hemophagocytic disorders?

- 1  yes
- 2  no
- 3  unknown

Specify affected member(s):

- 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)?

- 1  yes
- 2  no
- 3  unknown

9. Describe:

Specify the following clinical and laboratory features at diagnosis:

- |     | Present                    | Absent                     | Unknown                    |   |
|-----|----------------------------|----------------------------|----------------------------|---|
| 10. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Anemia (hemoglobin < 10 g/dL)                             |
| 11. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Fever (> 38.5° C for > 7 days within 1 week of diagnosis) |
| 12. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Hepatomegaly (> 3 cm below right costal margin)           |
| 13. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Hypertriglyceridemia (> 200 mg/dL)                        |
| 14. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Hypofibrinogenemia (< 150 mg/dL)                          |
| 15. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Neutropenia (ANC < 1.0 x 10 <sup>9</sup> /L)              |
| 16. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Splenomegaly (> 3 cm below left costal margin)            |
| 17. | 1 <input type="checkbox"/> | 2 <input type="checkbox"/> | 3 <input type="checkbox"/> | Thrombocytopenia (< 100 x 10 <sup>9</sup> /L)             |

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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:  :

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46. Other infection:

- 1  yes →  
2  no

47. Specify other infection:  
(see codes below)

48. If other organism (code 198, 209, 219, 259, 329, or 409), specify:  
\_\_\_\_\_

Specify infection site(s):

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 →  
54. 1  yes 2  no Other body fluid →

53. Specify tissue / site: \_\_\_\_\_

55. Specify fluid / site: \_\_\_\_\_

### Codes for Commonly Reported Organisms

Bacterial Infections		Fungal Infections		Viral Infections	
121 Acinetobacter	148 Leptotrichia buccalis	178 Streptococcus pneumoniae	200 Candida, NOS	301 Herpes simplex (HSV1, HSV2)	
122 Actinomyces	149 Leuconostoc (all species)	168 Treponema (syphilis)	201 Candida albicans	302 Varicella (herpes zoster, chicken pox)	
123 Bacillus	104 Listeria	169 Vibrio (all species)	206 Candida guilliermondii	303 Cytomegalovirus (CMV)	
124 Bacteroides (gracillis, uniformis, vulgaris, other species)	150 Methylobacterium	197 Multiple bacteria at a single site, specify bacterial codes	202 Candida krusei	304 Adenovirus	
125 Bordetella pertussis (whooping cough)	151 Micrococcus, NOS	198 Other bacteria, specify ‡	207 Candida lusitanae	305 Enterovirus (coxsackie, echo, polio)	
126 Borrelia (Lyme disease)	112 Mycobacterium avium–intracellulare (MAC, MAI)	501 Suspected atypical bacterial infection	203 Candida parapsilosis	306 Hepatitis A (HAV)	
127 Branhamella or Moraxella catarrhalis (other species)	174 Mycobacterium species (chelonae, fortuitum, haemophilum, kansasii, mucogenicum)	502 Suspected bacterial infection	204 Candida tropicalis	307 Hepatitis B (HBV, Australian antigen)	
128 Campylobacter (all species)	110 Mycobacterium tuberculosis (tuberculosis, Koch bacillus)		205 Candida (Torulopsis) glabrata	308 Hepatitis C (HCV)	
129 Capnocytophaga	175 Other mycobacterium, specify		209 Other Candida, specify ‡	309 HIV-1 (HTLV-III)	
171 Chlamydia pneumoniae	176 Mycobacterium, NOS		210 Aspergillus, NOS §	310 Influenza, NOS	
172 Other chlamydia, specify	105 Mycoplasma		211 Aspergillus flavus §	323 Influenza A	
113 Chlamydia, NOS	152 Neisseria (gonorrhoea, meningitidis, other species)		212 Aspergillus fumigatus §	324 Influenza B	
130 Citrobacter (freundii, other species)	106 Nocardia		213 Aspergillus niger §	311 Measles (rubeola)	
131 Clostridium (all species except difficile)	153 Pasteurella multocida		219 Other Aspergillus, specify ‡ §	312 Mumps	
132 Clostridium difficile	154 Propionibacterium (acnes, avidum, granulosum, other species)		220 Cryptococcus species	313 Progressive multifocal leukoencephalopathy (PML)	
173 Corynebacterium jeikeium	155 Proteus		230 Fusarium species §	314 Respiratory syncytial virus (RSV)	
133 Corynebacterium (all non-diphtheria species)	156 Pseudomonas (all species except cepacia & maltophilia)		261 Histoplasmosis	315 Rubella (German measles)	
101 Coxiella	157 Pseudomonas or Burkholderia cepacia		240 Zygomycetes, NOS §	316 Parainfluenza	
134 Enterobacter	158 Pseudomonas or Stenotrophomonas or Xanthomonas maltophilia		241 Mucormycosis §	317 Human herpesvirus-6 (HHV-6)	
177 Enterococcus, vancomycin resistant (VRE)	159 Rhodococcus		242 Rhizopus §	318 Epstein-Barr virus (EBV)	
135 Enterococcus (all species)	107 Rickettsia		250 Yeast, NOS	319 Polyoma virus (BK virus, JC virus)	
136 Escherichia (also E. coli)	160 Salmonella (all species)		259 Other fungus, specify ‡	320 Rotavirus	
137 Flavimonas oryzihabitans	161 Serratia marcescens		260 Pneumocystis (PCP / PJP)	321 Rhinovirus	
138 Flavobacterium	162 Shigella		503 Suspected fungal infection §	322 Human papilloma virus (HPV)	
139 Fusobacterium	163 Staphylococcus, coagulase negative (not aureus)			329 Other virus, specify ‡	
144 Haemophilus (all species, including influenzae)	164 Staphylococcus aureus			504 Suspected viral infection	
145 Helicobacter pylori	165 Staphylococcus, NOS				
146 Klebsiella	166 Stomatococcus mucilaginosus				
147 Lactobacillus (bulgaricus, acidophilus, other species)	167 Streptococcus (all species except Enterococcus)				
102 Legionella					
103 Leptospira					

§ For fungal species marked with a section symbol (§), also complete a Fungal Infection insert (SUP-FNG).

**Parasitic Infections**  
402 Toxoplasma  
403 Giardia  
404 Cryptosporidium  
409 Other parasite, specify ‡  
505 Suspected parasite infection

**Other Infections**  
509 No organism identified

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Specify the site(s) where hemophagocytosis was documented at diagnosis:

- |     | Present                  | Absent                   | Not tested               |              |
|-----|--------------------------|--------------------------|--------------------------|--------------|
| 56. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | Bone marrow  |
| 57. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | CSF          |
| 58. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | Liver        |
| 59. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | Lymph nodes  |
| 60. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | Spleen       |
| 61. | <input type="checkbox"/> | <input type="checkbox"/> | <input type="checkbox"/> | Other site → |

62. Specify site of hemophagocytosis:

63. What is the current natural killer cell function? (Refers to specific cytotoxicity 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?

- 1  yes →
- 2  no
- 3  unknown

Specify therapy and reason for therapy:

65. Cyclosporine:	<b>Given for</b> induction / maintenance?	<b>Given for disease relapse?</b>
1 <input type="checkbox"/> yes →	66. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	67. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no		
68. Intrathecal methotrexate:		
1 <input type="checkbox"/> yes →	69. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	70. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no		
71. IVIG:		
1 <input type="checkbox"/> yes →	72. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	73. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no		
74. Steroids:		
1 <input type="checkbox"/> yes →	75. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	76. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no		
77. VP-16 / VM-26:		
1 <input type="checkbox"/> yes →	78. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	79. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no		
80. Other drug:		
1 <input type="checkbox"/> yes →	81. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	82. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no	83. Specify other drug: _____	
84. Radiation therapy:		
1 <input type="checkbox"/> yes →	85. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no	86. 1 <input type="checkbox"/> yes 2 <input type="checkbox"/> no
2 <input type="checkbox"/> no	87. Specify radiation field: _____	
	88. Specify total dose: <input type="text"/> cGy	
89. Was this therapy given following the HLH-94 protocol of the Histiocyte Society?		
1 <input type="checkbox"/> yes		
2 <input type="checkbox"/> no		
3 <input type="checkbox"/> unknown		

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90. Was CNS disease quiescent (inactive) at any time prior to the preparative regimen?

- 1  yes
- 2  no
- 3  unknown

Specify:

- 91. 1  yes 2  no 3  unknown Normal or stable CT or MRI of CNS
- 92. 1  yes 2  no 3  unknown Normal CSF neopterin level
- 93. 1  yes 2  no 3  unknown Normal CSF protein
- 94. 1  yes 2  no 3  unknown Normal CSF WBC (< 5 cells/mm<sup>3</sup>)
- 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. 1  yes 2  no 3  unknown ANC > 1.0 x 10<sup>9</sup>/L (without growth factor support)
- 98. 1  yes 2  no 3  unknown Hemoglobin > 9 g/dL without transfusion
- 99. 1  yes 2  no 3  unknown Hepatomegaly resolved (≤ 3 cm below costal margin)
- 100. 1  yes 2  no 3  unknown Normal fibrinogen
- 101. 1  yes 2  no 3  unknown Normal triglycerides
- 102. 1  yes 2  no 3  unknown Platelets > 100 x 10<sup>9</sup>/L without transfusion
- 103. 1  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:

<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>
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:

not applicable

<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>
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:

not applicable

<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>	<input type="text"/>
Month		Day		Year	

110. Specify the site of the third relapse / reactivation:

- 1  CNS
- 2  systemic
- 3  both

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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<sup>9</sup>/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<sup>9</sup>/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: \_\_\_\_\_