National Marrow Donor Program®
Insert XVI – Hemophagocytic Lymphohistiocytosis

Registry Use Only

Sequence Number: ____________

Date Received: ____________

Unrelated

Recipient NMDP ID: ☐☐☐ - ☐☐☐ - ☐

Recipient Last Name: ☐☐☐☐☐☐☐☐☐☐☐☐☐☐☐

Recipient Local ID (optional): ☐☐☐☐☐☐☐☐☐☐

Today's Date: ☐☐ (Month) ☐☐ (Day) ☐☐☐☐ (Year) TC Code: ☐☐☐

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.

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1. Date of diagnosis: ☐☐ (Month) ☐☐ (Day) ☐☐☐☐ (Year)

2. Is there a family history of hemophagocytic disorders?
   - 1 ☐ yes → (go to question 3)
   - 2 ☐ no
   - 3 ☐ unknown

3. Specify affected member(s)
   - a. Sibling(s) 1 ☐ yes 2 ☐ no
   - b. Cousin(s) 1 ☐ yes 2 ☐ no
   - c. Aunt(s)/Uncle(s) 1 ☐ yes 2 ☐ no
   - d. Other 1 ☐ yes 2 ☐ no If yes, specify relationship: ____________

4. Is there a family history of consanguinity?
   - 1 ☐ yes → (go to question 5)
   - 2 ☐ no
   - 3 ☐ unknown

5. Describe: ____________

6. Clinical and laboratory features at time of diagnosis:

Feature			
a. Fever (> 38.5° C for > 7 days within 1 week of diagnosis)	1 ☐ present	2 ☐ absent	3 ☐ unknown
b. Splenomegaly (> 3 cm below left costal margin)	1 ☐ present	2 ☐ absent	3 ☐ unknown
c. Hepatomegaly(> 3 cm below right costal margin)	1 ☐ present	2 ☐ absent	3 ☐ unknown
d. Anemia (Hgb < 10 g/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown
e. Neutropenia (ANC < 1.0 x 10^9/L)	1 ☐ present	2 ☐ absent	3 ☐ unknown
f. Thrombocytopenia (< 100 x 10^9/L)	1 ☐ present	2 ☐ absent	3 ☐ unknown
g. Hypofibrinogenemia (< 150 mg/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown
h. Hypertriglyceridemia (> 200 mg/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown

7. Cerebrospinal fluid findings at time of diagnosis:

Finding			
a. WBC count:	1 ☐ ≤ 5 cells/µl	2 ☐ > 5 cells/µl	3 ☐ not tested
b. Neopterin level:	1 ☐ normal	2 ☐ elevated	3 ☐ not tested
c. Protein:	1 ☐ normal	2 ☐ elevated	3 ☐ not tested

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

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

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9. Were CNS abnormalities found on CT or MRI scans at any time prior to stem cell transplant conditioning?
1 ☐ yes → *(Please attach a copy of the report, and note that it pertains to question 9.)*
2 ☐ no
3 ☐ not tested

10. Type of scan:
1 ☐ CT
2 ☐ MRI

11. Specify abnormality: ________________

12. Were there any clinical neurologic abnormalities at the time of diagnosis or at any time prior to conditioning?
1 ☐ yes →
2 ☐ no
3 ☐ unknown

13. Specify:

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

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Infection History at Time of Presentation with Disease

14. Was an infection documented at the time of diagnosis?
1 ☐ yes →
2 ☐ no

15. EBV:
1 ☐ yes →
2 ☐ no

16. Diagnosis made by:
1 ☐ Serology →
2 ☐ PCR
3 ☐ In situ hybridization

17. Titers:
a. Viral capsid IgG: ☐ : ☐☐☐☐
b. Viral capsid IgM: ☐ : ☐☐☐☐
c. Early antigen: ☐ : ☐☐☐☐
d. EBNA: ☐ : ☐☐☐☐

18. CMV:
1 ☐ yes →
2 ☐ no

19. Diagnosis made by:
1 ☐ Serology →
2 ☐ PCR
3 ☐ Culture

20. Titers:
a. IgG: ☐ : ☐☐☐
b. IgM: ☐ : ☐☐☐

21. Other virus:
1 ☐ yes → 22. Specify: ________________
2 ☐ no

23. Other infection (bacterial, fungal, etc.):
1 ☐ yes → 24. Specify: ________________
2 ☐ no

25. From what site was the infection diagnosis made?

a. Blood	1 ☐ yes	2 ☐ no
b. Urine	1 ☐ yes	2 ☐ no
c. CSF	1 ☐ yes	2 ☐ no
d. Tissue biopsy, specify: ________	1 ☐ yes	2 ☐ no
e. Other body fluid, specify: ________	1 ☐ yes	2 ☐ no

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26. Indicate sites where hemophagocytosis was documented at the time of diagnosis:

a. Bone marrow	1 ☐ yes	2 ☐ no	3 ☐ not tested
b. CSF	1 ☐ yes	2 ☐ no	3 ☐ not tested
c. Spleen	1 ☐ yes	2 ☐ no	3 ☐ not tested
d. Liver	1 ☐ yes	2 ☐ no	3 ☐ not tested
e. Lymph node	1 ☐ yes	2 ☐ no	3 ☐ not tested
f. Other, specify site: ______________	1 ☐ yes	2 ☐ no	3 ☐ not tested

27. Natural killer cell function at time of diagnosis:
- 1 ☐ absent *(< 10% normal)*
- 2 ☐ decreased
- 3 ☐ normal
- 4 ☐ increased
- 5 ☐ not tested

28. Was treatment given at any time prior to conditioning?
- 1 ☐ yes → (go to question 29)
- 2 ☐ no
- 3 ☐ unknown

29. Specify therapy and reason for therapy:

		Induction/Maintenance	Relapse
a. VP-16 / VM-26	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
b. Steroids	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
c. IT methotrexate	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
d. Cyclosporin	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
e. IVIG	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
f. Other drug, specify: ______________	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no
g. Radiation therapy, specify field: ______________	1 ☐ no 2 ☐ yes →	1 ☐ yes 2 ☐ no	1 ☐ yes 2 ☐ no

Total dose: ☐☐☐☐ cGy

30. Was this therapy given following the HLH-94 protocol of the Histiocyte Society?
- 1 ☐ yes
- 2 ☐ no
- 3 ☐ unknown

31. Was CNS disease quiescent at any time prior to conditioning?
- 1 ☐ yes → (go to question 32)
- 2 ☐ no → (go to question 32)
- 3 ☐ unknown

32. Specify:

a. Normal CSF WBC (< 5 cells/mm^3)	1 ☐ yes	2 ☐ no	3 ☐ unknown
b. Normal CSF protein	1 ☐ yes	2 ☐ no	3 ☐ unknown
c. Normal CSF neopterin level	1 ☐ yes	2 ☐ no	3 ☐ unknown
d. Normal or stable CT or MRI of CNS	1 ☐ yes	2 ☐ no	3 ☐ unknown

(Please send copy of report and note that it pertains to question 32.)

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33. Was systemic disease quiescent at any time prior to conditioning?
   - 1 ☐ yes → 34
   - 2 ☐ no → 34
   - 3 ☐ unknown

34. Specify:

a. Hepatomegaly resolved (≤ 3 cm)	1 ☐ yes	2 ☐ no	3 ☐ unknown
b. Splenomegaly resolved (≤ 3 cm)	1 ☐ yes	2 ☐ no	3 ☐ unknown
c. Hgb > 9 g/dL without transfusion	1 ☐ yes	2 ☐ no	3 ☐ unknown
d. Platelets > 100 x 10^9/L without transfusion	1 ☐ yes	2 ☐ no	3 ☐ unknown
e. ANC > 1.0 x 10^9/L	1 ☐ yes	2 ☐ no	3 ☐ unknown
f. Normal fibrinogen	1 ☐ yes	2 ☐ no	3 ☐ unknown
g. Normal triglycerides	1 ☐ yes	2 ☐ no	3 ☐ unknown

35. Were there any signs of relapse/reactivation prior to BMT?
   - 1 ☐ yes → 36
   - 2 ☐ no

36. Date:	☐☐ Month ☐☐ Day ☐☐☐☐ Year	1 ☐ systemic	2 ☐ CNS	3 ☐ both
37. Date:	☐☐ Month ☐☐ Day ☐☐☐☐ Year	1 ☐ systemic	2 ☐ CNS	3 ☐ both
38. Date:	☐☐ Month ☐☐ Day ☐☐☐☐ Year	1 ☐ systemic	2 ☐ CNS	3 ☐ both

Report treatments for relapse/reactivation in question 29.

39. Clinical and laboratory features just prior to conditioning:

a. Fever (> 38.5° C for > 7 days within 1 week of conditioning)	1 ☐ present	2 ☐ absent	3 ☐ unknown
b. Splenomegaly (> 3 cm below left costal margin)	1 ☐ present	2 ☐ absent	3 ☐ unknown
c. Hepatomegaly (> 3 cm below right costal margin)	1 ☐ present	2 ☐ absent	3 ☐ unknown
d. Anemia (Hgb < 10 g/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown
e. Neutropenia (ANC < 1.0 x 10^9/L)	1 ☐ present	2 ☐ absent	3 ☐ unknown
f. Thrombocytopenia (< 100 x 10^9/L)	1 ☐ present	2 ☐ absent	3 ☐ unknown
g. Hypofibrinogenemia (< 150 mg/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown
h. Hypertriglyceridemia (> 200 mg/dL)	1 ☐ present	2 ☐ absent	3 ☐ unknown

40. Status of central nervous system disease just prior to conditioning: *(Please see instructions for definitions.)*
   - 1 ☐ Active
   - 2 ☐ Non-active, quiescent
   - 3 ☐ CNS disease absent at diagnosis

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