Form 2156 R1.0: Pigmentary Dilution Disorder (PDD) Post-HCT Data

Center: CRID:

Key Fields

Sequence Number: _____

Date Received: __ __ __ __ - __ __- __ __

CIBMTR Center Number: _____

CIBMTR Recipient ID: _____

Date of HCT for which this form is being completed: __ __ __ __ - __ __- __ __

HCT Type: (check all that apply)

- Autologous
- Allogeneic, unrelated
- Allogeneic, related

Product type: (check all that apply)

- Bone marrow
- PBSC
- Single cord blood unit
- Multiple cord blood units
- Other product

Specify: _____

Visit

100 day 6 months 1 year 2 years > 2 years,

Specify: _____

Disease Assessment Since the Date of Last Report

Questions: 1 - 24

Indicate which of the following manifestations of the disease were present since the date of the last report:

1 Giant leukocyte granules

yes no Unknown

2 Neutropenia (ANC < 1.0 x 10^9 /L)

yes no Unknown

3 Recurrent infections

yes no Unknown

4 Thrombocytopenia (platelets < 100 x 10^9 /L)

yes no Unknown

5 Bleeding diathesis

yes no Unknown

6 Bleeding from the GI tract

yes no

7 Easy bruising

yes no

8 Hematuria

yes no

9 Oral bleeding

yes no

10 Recurrent nosebleeds

yes no

11 Other bleeding

yes no

12 Specify other bleeding: _____

13 Did any clinical neurologic abnormalities persist or develop?

yes no Unknown

Specify neurologic dysfunction(s):

14 Abnormal gait

yes no

15 Areflexia

yes no

16 Ataxia and/or other symptoms of cerebellar dysfunction

yes no

17 Developmental delay

yes no

18 Was the recipient's IQ tested?

yes no

19 Date IQ was tested: __ __ __ __ - __ __- __ __

20 IQ test instrument

Kaufman Assessment Battery for Children

Raven's Progressive Matrices

Stanford-Binet

Wechsler Adult Intelligence Scale

Wechsler Intelligence Scale for Children

Woodcock-Johnson Tests of Cognitive Abilities

21 IQ score: _____

22 Seizures

yes no

23 Other neurologic abnormality

yes no

24 Specify other neurologic abnormality: _____

Accelerated Phase (AP)

Questions: 25 - 51

25 Did the recipient develop features of an accelerated phase since the date of the last report?

yes no Unknown

26 Number of accelerated phases recorded: _____

27 Date first accelerated phase detected
(in this reporting period)

Known Unknown

28 Date first accelerated phase was detected: __ __ __ __ - __ __- __ __

29 Abnormal CSF
(WBC >5 cells/μL, elevated protein)
yes no Unknown

30 Abnormal liver function
yes no Unknown

31 Anemia
(Hgb <10 g/dL)
yes no Unknown

32 Cytomegalovirus (CMV)
(associated with accelerated phase)
yes no Unknown

33 Epstein-Barr virus (EBV)
(associated with accelerated phase)
yes no Unknown

34 Other viral infection associated with accelerated phase
(not CMV or EBV)
yes no Unknown

35 Specify other infection: _____

36 Dense bodies (delta granules) on electron micrograph (EM) of platelets
yes no Unknown

37 Elevated triglycerides
yes no Unknown

38 Fevers
(> 38.5° C or > 101.3° F for > 7 days)
yes no Unknown

39 Hemophagocytosis
yes no Unknown

40 Hepatomegaly (liver edge palpable > 3 cm below right costal margin)
yes no Unknown

41 Hyperferritinemia
(serum ferritin > 500 ng/mL or > 500 μg/dL)
yes no Unknown

42 Hypofibrinogenemia
(serum fibrinogen < 150 mg/dL or < 1.5 g/L or < 4.4 μmol/L)
yes no Unknown

43 Hyponatremia
(serum sodium < 135 mg/dL)
yes no Unknown

44 Lymphadenopathy
yes no Unknown

45 CSF lymphocytosis
yes no Unknown

46 Neurologic dysfunction
(e.g. seizures, meningitis signs)
yes no Unknown

47 Neutropenia (ANC < 1.0 x 10^9 /L)
yes no Unknown

48 Splenomegaly (spleen palpable > 3 cm below left costal margin)
yes no Unknown

49 Thrombocytopenia (platelets < 100 x 10^9 /L)
yes no Unknown

50 Other feature associated with accelerated phase
yes no Unknown

51 Specify other feature: _____

Post-HCT Therapy

Questions: 52 - 64

52 Was therapy given?
yes no Unknown

Specify therapy given:

53 Acyclovir
yes no

54 Antithymocyte globulin (ATG)
yes no

55 Corticosteroids
yes no

56 Cyclosporine (CSA, Neoral, Sandimmune)
yes no

57 Etoposide (VP-16, VePesid)
yes no

58 Ganciclovir (DHPG)
yes no

59 Intrathecal methotrexate (IT MTX)
yes no

60 Intravenous immune globulin (IVIG)
yes no

61 Interferon-α (Intron, Roferon) (includes PEG)
yes no

62 Rituximab (Rituxan, MabThera)
yes no

63 Other therapy
yes no

64 Specify other therapy: _____

Current Assessment of Immunologic Function Post-HCT

Questions: 65 - 82

65 Cytotoxic T-cell activity

- Absent (< 10% of control)
- Low (10–30% of control)
- Normal (> 30% of control)
- Not done

66 Date sample collected

- Known
- Unknown

67 Date sample collected: __ __ __ __ - __ __- __ __

68 Degranulation of cytolytic lymphocytes (CD107a expression)

- Absent (< 10% of control)
- Low (10–30% of control)
- Normal (> 30% of control)
- Not done

69 Date sample collected

- Known
- Unknown

70 Date sample collected: __ __ __ __ - __ __- __ __

71 Granulocyte chemotaxis

- Absent (< 10% of control)
- Low (10-30% of control)
- Normal (>30% of control)
- Not done

72 Date sample collected

- Known
- Unknown

73 Date sample collected: __ __ __ __ - __ __- __ __

74 Natural killer cell activity
(against K562 cells)

- Absent (<10% of control)
- Low (10-30% of control)
- Normal (>30% of control)
- Not done

75 Date sample collected

- Known
- Unknown

76 Date sample collected: __ __ __ __ - __ __- __ __

77 Phytohemagglutinin (PHA)

- Absent (<10% of control)
- Low (10-30% of control)
- Normal (>30% of control)
- Not done

78 Date sample collected

- Known
- Unknown

79 Date sample collected: __ __ __ __ - __ __- __ __

80 Platelet aggregation

- Absent (<10% of control)
- Low (10-30% of control)
- Normal (>30% of control)
- Not done

81 Date sample collected

- Known
- Unknown

82 Date sample collected: __ __ __ __ - __ __- __ __

Disease Status at the Time of Evaluation for This Reporting Period

Questions: 83 - 84

83 What is the current disease status?

- No prior accelerated phase
- In remission from accelerated phase
- In accelerated phase
- Unknown

84 Date assessed: __ __ __ __ - __ __- __ __

First Name: _____

Last Name: _____

E-mail address: _____

Date: __ __ __ __ - __ __- __ __