Form 2055 R2.0: Chronic Granulomatous Disease (CGD) Pre-HSCT Data

Center: CRID:

Key Fields

Sequence Number: _____

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

CIBMTR Center Number: _____

CIBMTR Recipient ID: _____

Has this patient's data been previously reported to USIDNET?

yes no

USIDNET ID: _____

Today's Date: __ __ __ __ - __ __- __ __

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

HSCT type (check all that apply):

Autologous

Allogeneic, unrelated

Allogeneic, related

Syngeneic (identical twin)

Product type (check all that apply):

Marrow

PBSC

Cord blood

Other product

Specify: _____

If this is a report of a second or subsequent transplant, check here and continue with question 107.

Disease Assessment at Diagnosis

Questions: 1 - 6

1 What was the date of diagnosis of Chronic Granulomatous Disease (CGD)? __ __ __ __ - __ __- __ __

2 What is the pattern of CGD inheritance?

sporadic (no family history)

x-linked, documented

autosomal recessive, documented

Unknown

3 Are the parents of the patient consanguineous (related by blood ancestry)?

yes no Unknown

4 Are there other blood relatives in the patient's family with immunodeficiency disease?

yes no Unknown

5 What is the CGD molecular abnormality?

X-linked (gp91)

autosomal recessive - $p22^{phox}$

autosomal recessive - $p47^{phox}$

autosomal recessive - $p67^{phox}$

Unknown

6 Was a mutated protein / enzyme expressed?

yes no Unknown

Laboratory Studies at Diagnosis

Questions: 7 - 33

Report findings prior to any first treatment of chronic granulomatous disease.

7 Date CBC tested: (testing done within 6 weeks of diagnosis) __ __ __ __ - __ __- __ __

8 WBC: _____

x 10^9/L (x 10^3/mm^3)

x 10^6/L

WBC not tested

9 Lymphocytes: _____ Lymphocytes not tested

10 Eosinophils: _____ Eosinophils not tested

11 Polymorphonuclear leukocytes (PMN): _____ Polymorphonuclear leukocytes (PMN) not tested

12 Hemoglobin: _____

g/dL g/L mmol/L

Hemoglobin not tested

transfused RBC < 30 days from date of test

13 Platelets: _____

x 10^9/L (x 10^3/mm^3)

x 10^6/L

Platelets not tested

transfused platelets < 7 days from date of test

Lymphocyte Analysis

Specify the following lymphocyte analyses performed prior to any disease treatment:

14 Were lymphocyte analyses performed?

yes no

15 Date of most recent testing performed: __ __ __ __ - __ __- __ __

16 Absolute lymphocyte count: _____ cells / µL (cells / mm^3)

17 CD3 (T cells) % of total lymphocytes _____ %

-- or --

CD3 (T cells) value _____

x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD3 (T cells) not tested

18 CD4 (T helper cells) % of total lymphocytes _____ %

-- or --

CD4 (T helper cells) value _____

x 10^9/L (x 10^3/mm^3)

x 10^6/L

CD4 (T helper cells) not tested

19 CD8 (cytotoxic T cells) % of total lymphocytes _____ %

-- or --

CD8 (cytotoxic T cells) value _____

- x 10^9/L (x 10^3/mm^3)
- x 10^6/L

- CD8 (cytotoxic T cells) not tested

20 CD20 (B lymphocyte cells) % of total lymphocytes _____ %

-- or --

CD20 (B lymphocyte cells) value _____

- x 10^9/L (x 10^3/mm^3)
- x 10^6/L

- CD20 (B lymphocyte cells) not tested

21 CD56 (natural killer (NK) cells) % of total lymphocytes _____ %

-- or --

CD56 (natural killer (NK) cells) value _____

- x 10^9/L (x 10^3/mm^3)
- x 10^6/L

- CD56 (natural killer (NK) cells) not tested

22 CD4+/CD45RA+ (naive T cells) % of total lymphocytes _____ %

-- or --

CD4+/CD45RA+ (naive T cells) value _____

- x 10^9/L (x 10^3/mm^3)
- x 10^6/L

- CD4+ / CD45RA+ (naive T cells) not tested

23 CD4+/CD45RO+ (memory T cells) % of total lymphocytes _____ %

-- or --

CD4+/CD45RO+ (memory T cells) value _____

- x 10^9/L (x 10^3/mm^3)
- x 10^6/L

- CD4+/CD45RO+ (memory T cells) not tested

WBC Functional Assays

Specify the following WBC functional assays performed prior to any disease treatment:

24 Bacterial susceptibility testing/bacterial killing/drug resistance testing

- Normal
- Abnormal
- Not tested

25 Dichlorofluorescein (DCF)

- Normal
- deficient
- Not tested

26 Dihydrorhodamine oxidation (DHR)

- Normal
- deficient
- Not tested

27 Specify stimulation index: _____

- Unknown

28 Hydrogen peroxide production

Normal absent / decreased Not tested

29 Nitroblue tetrazolium test (NBT)

Normal Absent Not tested

30 Specify method:

slide (histochemical)

quantitative

Unknown

31 Superoxide production

Normal absent / decreased Not tested

32 Other lab test result

Normal Abnormal Not tested

33 Specify other test: _____

Clinical Features Assessed between Diagnosis and the Start of the Preparative Regimen

Questions: 34 - 132

34 Site of infection: adenitis

yes no

Adenitis (1)

Questions: 35 - 36

35 Organism _____

36 Specify other organism _____

37 If adenitis was present, was it a prominent feature of CGD?

yes no

38 Site of infection: brain abscess

yes no

Brain Abscess (1)

Questions: 39 - 40

39 Organism _____

40 Specify other organism _____

41 If brain abscess was present, was it a prominent feature of CGD?

yes no

42 Site of infection: cellulitis

yes no

Cellulitis (1)

Questions: 43 - 44

43 Organism _____

44 Specify other organism _____

45 If cellulitis was present, was it a prominent feature of CGD?

yes no

46 Site of infection: furuncles

yes no

Furuncles (1) **Questions: 47 - 48**

47 Organism _____

48 Specify other organism _____

49 If furuncles were present, was it a prominent feature of CGD?

yes no

50 Site of infection: genitourinary

yes no

Genitourinary (1) **Questions: 51 - 52**

51 Organism _____

52 Specify other organism _____

53 If genitourinary infection was present, was it a prominent feature of CGD?

yes no

54 Site of infection: impetigo

yes no

Impetigo (1) **Questions: 55 - 56**

55 Organism _____

56 Specify other organism _____

57 If impetigo was present, was it a prominent feature of CGD?

yes no

58 Site of infection: joint

yes no

Joint (1) **Questions: 59 - 60**

59 Organism _____

60 Specify other organism _____

61 If joint infection was present, was it a prominent feature of CGD?

yes no

62 Site of infection: liver abscess

yes no

Liver Abscess (1) **Questions: 63 - 64**

63 Organism _____

64 Specify other organism _____

65 If liver abscess was present, was it a prominent feature of CGD?

yes no

66 Site of infection: lung abscess

yes no

Lung Abscess (1) **Questions: 67 - 68**

67 Organism _____

68 Specify other organism _____

69 If lung abscess was present, was it a prominent feature of CGD?

yes no

70 Site of infection: lymph nodes abscess

yes no

Lymph Nodes Abscess (1) **Questions: 71 - 72**

71 Organism _____

72 Specify other organism _____

73 If lymph nodes abscess was present, was it a prominent feature of CGD?

yes no

74 Site of infection: meningitis / encephalitis

yes no

Meningitis / Encephalitis (1) **Questions: 75 - 76**

75 Organism _____

76 Specify other organism _____

77 If meningitis / encephalitis was present, was it a prominent feature of CGD?

yes no

78 Site of infection: osteomyelitis

yes no

Osteomyelitis (1) **Questions: 79 - 80**

79 Organism _____

80 Specify other organism _____

81 If osteomyelitis was present, was it a prominent feature of CGD?

yes no

82 Site of infection: perirectal abscess

yes no

Perirectal Abscess (1) **Questions: 83 - 84**

83 Organism _____

84 Specify other organism _____

85 If perirectal abscess was present, was it a prominent feature of CGD?

yes no

86 Site of infection: pneumonia

yes no

Pneumonia (1) **Questions: 87 - 88**

87 Organism _____

88 Specify other organism _____

89 If pneumonia was present, was it a prominent feature of CGD?

yes no

90 Site of infection: severe or protracted diarrhea

yes no

Severe or Protracted Diarrhea (1) **Questions: 91 - 92**

91 Organism _____

92 Specify other organism _____

93 If severe or protracted diarrhea was present, was it a prominent feature of CGD?

yes no

94 Site of infection: subcutaneous abscess

yes no

Subcutaneous Abscess (1) **Questions: 95 - 96**

95 Organism _____

96 Specify other organism _____

97 If subcutaneous abscess was present, was it a prominent feature of CGD?

yes no

98 Site of infection: systemic infection

yes no

Systemic Infection (1) **Questions: 99 - 100**

99 Organism _____

100 Specify other organism _____

101 If systemic infection was present, was it a prominent feature of CGD?

yes no

102 Site of infection: other infection

yes no

Other Infection (1) **Questions: 103 - 105**

103 Organism _____

104 Specify other organism _____

105 Specify other infection site: _____

106 If other infection was present, was it a prominent feature of CGD?

yes no

Clinical Status between Diagnosis and the Preparative Regimen

107 Did the recipient experience any of the following clinical features (between diagnosis and prior to the preparative regimen)?

yes no

108 Is autoimmune hemolytic anemia present?

yes no

109 Is autoimmune hemolytic anemia prominent?

yes no

110 Was autoimmune hemolytic anemia also present at the time of first treatment for CGD?

yes no

111 Is failure to thrive (weight<5th percentile) present?

yes no

112 Is failure to thrive (weight < 5th percentile) prominent?

yes no

113 Was failure to thrive (weight < 5th percentile) also present at the time of first treatment fo CGD?

yes no

114 Is gastric outlet obstruction present?

yes no

115 Is gastric outlet obstruction prominent?

yes no

116 Was gastric outlet obstruction also present at the time of first treatment for CGD?

yes no

117 Is growth retardation (height<5th percentile) present?

yes no

118 Is growth retardation (height < 5th percentile) prominent?

yes no

119 Was growth retardation (height < 5th percentile) also present at the time of first treatment for CGD?

yes no

120 Is inflammatory bowel disease present?

yes no

121 Is inflammatory bowel disease prominent?

yes no

122 Was inflammatory bowel disease also present at the time of first treatment for CGD?

yes no

123 Is thrombocytopenia (< 100 x 10^9/L) present?

yes no

124 Is thrombocytopenia (< 100 x 10^9/L) prominent?

yes no

125 Was thrombocytopenia (< 100 x 10^9/L) also present at the time of first treatment for CGD?

yes no

126 Is urinary outlet obstruction present?

yes no

127 Is urinary outlet obstruction prominent?

yes no

128 Was urinary outlet obstruction also present at the time of first treatment for CGD?

yes no

129 Are other clinical features present?

yes no

130 Are other clinical features prominent?

yes no

131 Were other clinical features also present at the time of first treatment for CGD?

yes no

132 Specify other clinical feature: _____

Pre-HSCT Treatment for Chronic Granulomatous Disease

Questions: 133 - 172

133 Was treatment given (between diagnosis and prior to the preparative regimen)?

yes no

Prophylactic drugs paused for < 1 week should *not* be considered as "Prophylactic Drug Stopped."

134 Were antifungal drug(s) given as prophylaxis?

yes no

135 Were prophlactic antifungal drug(s) stopped?

yes no

136 Date prophylactic antifungal drug(s) stopped unknown

date estimated

Date prophylactic antifungal drug(s) stopped: __ __ __ __ - __ __- __ __

137 Was co-trimoxazole (Bactim, Septra) given as prophylaxis?

yes no

138 Was co-trimoxazole (Bactrim, Septra) stopped?

yes no

139 Date co-trimoxazole (Bactrim, Septra) stopped unknown

date estimated

Date co-trimoxazole (Bactrim, Septra) stopped: __ __ __ __ - __ __- __ __

140 Was interferon-gamma (immune interferon, IFN-g) given as prophylaxis?

yes no

141 Was interferon-gamma (immune interferon, IFN-g) stopped?

yes no

142 Date interferon-gamma (immune interferon, IFN-g) stopped unknown

date estimated

Date interferon-gamma (immune interferon, IFN-g) stopped: __ __ __ __ - __ __- __ __

Therapy paused for < 1 week should not be considered as "Therapy Stopped."

143 Were systemic corticosteroids given as therapy?

yes no

144 Were systemic corticosteroids stopped?

yes no

145 Date therapeutic systemic corticosteroids stopped unknown

date estimated

Date systemic corticosteroids stopped: __ __ __ __ - __ __- __ __

146 Was other immunosuppressive drug given as therapy?

yes no

147 Was other immunosuppressive drug stopped?

yes no

148 Date other therapeutic immunosuppressive drug stopped unknown

date estimated

Date therapeutic other other immunosuppressive drugs stopped: __ __ __ __ - __ __- __ __

149 Specify other immunosuppressive drug: _____

150 Was gene therapy performed (between diagnosis and prior to the preparative regimen)?

yes no

151 Specify date of infusion of gene therapy: __ __ __ __ - __ __- __ __

152 Was the recipient considered to have failed gene therapy?

yes no

153 Did the recipient receive any other significant treatment(s) (between diagnosis and prior to the preparative regimen)?

yes no

154 Specify other treatment(s): _____

155 Did the recipient receive parenteral nutrition (between diagnosis and prior to the preparative regimen)?

yes no

156 Was parenteral nutrition in use a the time of transplantation?

yes no

157 Did the recipient receive mechanical ventilation (between diagnosis and prior to the preparative regimen)?

yes no

158 Was mechanical ventilation in use at the time of transplantation?

yes no

159 Were any biologic specimens collected for this recipient (between diagnosis and prior to the preparative regimen)?

yes no Unknown

Specify specimen(s) collected and available for future research:

160 DNA

yes no

161 Epstein-Barr virus (EBV)-transformed B-Cell line

yes no

162 Fibroblast cell line

yes no

163 Herpes virus saimiri-transformed T-cell line

yes no

164 Other T-cell line

yes no

165 Pathological specimen

yes no

166 Specify pathological speciment(s): _____

167 Peripheral blood mononuclear cells (PBMC), frozen

yes no

168 RNA

yes no

169 Specify RNA source: _____

170 Serum (pre-IVIG)

yes no

171 Other specimen

yes no

172 Specify other biologic specimen(s): _____

First Name: _____ Last Name: _____

Phone number: _____ Fax number: _____

E-mail address: _____