Form 2044 R2.0: Systemic Sclerosis Pre-HSCT Data

Center: CRID:

Key Fields

Sequence Number: _____

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

CIBMTR Center Number: _____

CIBMTR Recipient 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 180.

Disease Assessment at Diagnosis   Questions: 1 - 2

1 What was the date of diagnosis of Systemic Sclerosis (SSc)? __ __ __ __ - __ __- __ __

2 What was the extent of cutaneous systemic sclerosis at diagnosis?

☐ limited (cutaneous thickening distal (but not proximal) to elbows or knees)

☐ diffuse

☐ Unknown

Laboratory Studies at Diagnosis   Questions: 3 - 29

Report findings prior to any first treatment for systemic sclerosis.

3 Date CBC tested: __ __ __ __ - __ __- __ __

4 WBC:

☐ Known   ☐ Not known

5 _____

☐ $x\ 10^9/L\ (x\ 10^3/mm^3)$

☐ $x\ 10^6/L$

6 Hemoglobin:

☐ Known   ☐ Not known

7 _____

☐ g/dL   ☐ g/L   ☐ mmol/L

8 Hematocrit:

☐ Known   ☐ Not known

9 _____ %

10 Platelets:

☐ Known   ☐ Not known

11 _____   ☐ x 10^9/L (x 10^3/mm^3)

☐ x 10^6/L

12 Serum creatinine:

☐ Known   ☐ Not known

13 _____   ☐ mg/dL   ☐ mmol/L   ☐ µmol/L

14 Creatinine clearance:

☐ Known   ☐ Not known

15 _____   ☐ mL/min   ☐ mL/sec

16 Creatinine phosphokinase:

☐ Known   ☐ Not known

17 _____   ☐ U/L   ☐ µkat/L

18 Blood urea nitrogen:

☐ Known   ☐ Not known

19 _____   ☐ mg/dL   ☐ mmol/L

20 Was there evidence of hematuria at diagnosis?

☐ yes, present   ☐ no, absent   ☐ Unknown

21 Was there evidence of proteinuria at diagnosis?

☐ yes, present   ☐ no, absent   ☐ Unknown

22 Thyroid stimulating hormone (TSH):

☐ Known   ☐ Not known

23 _____   ☐ mU/L   ☐ µU/ml

24 Was any testing for autoantibodies performed at diagnosis?

☐ yes   ☐ no   ☐ Unknown

Specify the test results for the following autoantibodies:

25 Anti-centromere:

☐ Positive   ☐ Negative   ☐ Inconclusive   ☐ not tested / unknown

26 Anti-DNA topoi-
somerase I (Scl-70):

☐ Positive   ☐ Negative   ☐ Inconclusive   ☐ not tested / unknown

27 Anti-nuclear:

☐ Positive   ☐ Negative   ☐ Inconclusive   ☐ not tested / unknown

28 Anti-SS-A:

☐ Positive   ☐ Negative   ☐ Inconclusive   ☐ not tested / unknown

29 Anti-SS-B:

     ☐ Positive   ☐ Negative   ☐ Inconclusive   ☐ not tested / unknown

Pre-HSCT Treatment for Systemic Lupus Erythematosus	Questions: 30 - 81

30 Did the recipient receive any disease-modifying treatments between the time of diagnosis and prior to mobilization for stem cell collection (or high dose therapy if mobilization was not done)?

     ☐ yes   ☐ no

31 Antithymocyte globulin (ATG) / antilymphocyte globulin (ALG)

     ☐ yes   ☐ no   ☐ Unknown

     32 Treatment Stopped

         ☐ yes   ☐ no

         33 Stopped Code: _____

            34 Specify: _____

35 Cyclophosphamide (CTX, Cytoxan, Neosar)

     ☐ yes   ☐ no   ☐ Unknown

     36 Treatment Stopped

         ☐ yes   ☐ no

         37 Stopped Code: _____

            38 Specify: _____

39 Cyclosporine (CsA, Neoral, Sandimmune)

     ☐ yes   ☐ no   ☐ Unknown

     40 Treatment Stopped

         ☐ yes   ☐ no

         41 Stopped Code: _____

            42 Specify: _____

43 D-penicillamine (penicillamine, Cuprimine, Depen)

     ☐ yes   ☐ no   ☐ Unknown

     44 Treatment Stopped

         ☐ yes   ☐ no

         45 Stopped Code: _____

            46 Specify: _____

47 Methotrexate (MTX, Folex)

     ☐ yes   ☐ no   ☐ Unknown

     48 Treatment Stopped

         ☐ yes   ☐ no

         49 Stopped Code: _____

            50 Specify: _____

     51 ☐ Maximum weekly dose unknown:

Specify the maximum weekly dose: _____ mg

     52 ☐ Duration of therapy unknown:

Specify the duration of therapy: _____ ☐ weeks   ☐ months

53 Mycophenolate Mofetil (MMF, CellCept)

   ☐ yes   ☐ no   ☐ Unknown

> **54** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **55** Stopped Code: _____
> >
> >   **56** Specify: _____

57 Non-steroidal anti-inflammatory drugs (NSAIDS)

   ☐ yes   ☐ no   ☐ Unknown

> **58** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **59** Stopped Code: _____
> >
> >   **60** Specify: _____

61 Phototherapy

   ☐ yes   ☐ no   ☐ Unknown

> **62** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **63** Stopped Code: _____
> >
> >   **64** Specify: _____

65 Prednisone (Intensol, Sterapred) or equivalent

   ☐ yes   ☐ no   ☐ Unknown

> **66** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **67** Stopped Code: _____
> >
> >   **68** Specify: _____

69 Prostanoids / prostaglandin analogs

   ☐ yes   ☐ no   ☐ Unknown

> **70** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **71** Stopped Code: _____
> >
> >   **72** Specify: _____

73 Tacrolimus (FK 506, Prograf)

   ☐ yes   ☐ no   ☐ Unknown

> **74** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **75** Stopped Code: _____
> >
> >   **76** Specify: _____

77 Other treatment

   ☐ yes   ☐ no   ☐ Unknown

> **78** Treatment Stopped
>
>   ☐ yes   ☐ no
>
> > **79** Stopped Code: _____

80   Specify: _____

81   Specify other treatment: _____

Disease Assessment Prior to Mobilization Therapy for Stem Cell Collection   Questions: 82 - 114

Information for this section should come from the most recent evaluation prior to the initiation of mobilization therapy (<= 4 weeks prior to mobilization for stem cell collection). If the recipient did not receive mobilization therapy, check here and continue with question 180.

82   Date of evaluation prior to mobilization for stem cell collection: __ __ __ __ - __ __ - __ __

83   What was the extent of cutaneous systemic sclerosis prior to mobilization for stem cell collection?

    ○   limited (cutaneous thickening distal (but not proximal) to elbows or knees)

    ○   diffuse

    ○   Unknown

Specify skin thickness for the following sites as determined by clinical palpation performed prior to mobilization for stem cell collection: *Clements P, Lachenbruch P, Seibold J, White B, Weiner S, Martin R, et al. Inter and intraobserver variability of total skin thickness score (modified Rodnan TSS) in systemic sclerosis. J Rheumatol 1995; 22:1281-1285.*

84   Face

    ○   score 0   - Normal

    ○   score 1   - Mild

    ○   score 2   - Moderate

    ○   score 3   - Severe (inability to pinch skin into a fold)

85   Anterior chest

    ○   score 0   - Normal

    ○   score 1   - Mild

    ○   score 2   - Moderate

    ○   score 3   - Severe (inability to pinch skin into a fold)

86   Abdomen

    ○   score 0   - Normal

    ○   score 1   - Mild

    ○   score 2   - Moderate

    ○   score 3   - Severe (inability to pinch skin into a fold)

87   Upper arm - left

    ○   score 0   - Normal

    ○   score 1   - Mild

    ○   score 2   - Moderate

    ○   score 3   - Severe (inability to pinch skin into a fold)

88   Upper arm - right

    ○   score 0   - Normal

    ○   score 1   - Mild

    ○   score 2   - Moderate

    ○   score 3   - Severe (inability to pinch skin into a fold)

89 Forearm-left

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

90 Forearm-right

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

91 Dorsum of hand - left

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

92 Dorsum of hand - right

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

93 Fingers - left

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

94 Fingers - right

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

95 Thigh - left

     score 0   - Normal

     score 1   - Mild

     score 2   - Moderate

     score 3   - Severe (inability to pinch skin into a fold)

96   Thigh - right

    score 0   - Normal

    score 1   - Mild

    score 2   - Moderate

    score 3   - Severe (inability to pinch skin into a fold)

97   Lower leg - left

    score 0   - Normal

    score 1   - Mild

    score 2   - Moderate

    score 3   - Severe (inability to pinch skin into a fold)

98   Lower leg - right

    score 0   - Normal

    score 1   - Mild

    score 2   - Moderate

    score 3   - Severe (inability to pinch skin into a fold)

99   Dorsum of foot - left

    score 0   - Normal

    score 1   - Mild

    score 2   - Moderate

    score 3   - Severe (inability to pinch skin into a fold)

100   Dorsum of foot - right

    score 0   - Normal

    score 1   - Mild

    score 2   - Moderate

    score 3   - Severe (inability to pinch skin into a fold)

101   Total modified Rodnan Skin Score: (add scores from questions 84-100) _____

Specify the following clinical findings prior to mobilization for stem cell collection:

102   Changes in skin pigmentation:

    Present   Absent   Unknown

103   Raynaud's phenomenon:

    Present   Absent   Unknown

104   Painful digital ulcers:

    Present   Absent   Unknown

    105   Specify number of digital ulcers: _____

106   Gut dysmotility

    Present   Absent   Unknown

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Center: _____   CRID: _____

107 Malabsorption:

⬡ Present ⬡ Absent ⬡ Unknown

108 Weight loss > 10% of body weight:

⬡ Present ⬡ Absent ⬡ Unknown

109 Muscle weakness:

⬡ Present ⬡ Absent ⬡ Unknown

110 Joint tenderness:

⬡ Present ⬡ Absent ⬡ Unknown

111 Specify number of joints affected: _____

112 Tendon friction rubs:

⬡ Present ⬡ Absent ⬡ Unknown

113 Specify number of sites affected: _____

114 Contractures:

⬡ Present ⬡ Absent ⬡ Unknown

Laboratory Studies Prior to Mobilization for Stem Cell Collection	Questions: 115 - 179

115 Date CBC tested: __ __ __ __ - __ __ - __ __

116 WBC:

⬡ Known ⬡ Not known

117 _____ ⬡ $x\ 10^9$/L ($x\ 10^3$/mm^3)

⬡ $x\ 10^6$/L

118 Hemoglobin:

⬡ Known ⬡ Not known

119 _____ ⬡ g/dL ⬡ g/L ⬡ mmol/L

120 Hematocrit:

⬡ Known ⬡ Not known

121 _____ %

122 Platelets:

⬡ Known ⬡ Not known

123 _____ ⬡ $x\ 10^9$/L ($x\ 10^3$/mm^3)

⬡ $x\ 10^6$/L

124 Serum creatinine:

⬡ Known ⬡ Not known

125 _____ ⬡ mg/dL ⬡ mmol/L ⬡ µmol/L

126 Creatinine clearance:

⬡ Known ⬡ Not known

127 _____ ⬡ mL/min ⬡ mL/sec

128 Creatinine phosphokinase:

  ⬚ Known   ⬚ Not known

   129 _____   ⬚ U/L   ⬚ µkat/L

130 Blood urea nitrogen:

  ⬚ Known   ⬚ Not known

   131 _____   ⬚ mg/dL   ⬚ mmol/L

132 Was there evidence of hematuria prior to mobilization therapy for stem cell collection?

  ⬚ yes, present   ⬚ no, absent   ⬚ Unknown

133 Was there evidence of proteinuria prior to mobilization therapy for stem cell collection?

  ⬚ yes, present   ⬚ no, absent   ⬚ Unknown

134 Thyroid stimulating hormone (TSH):

  ⬚ Known   ⬚ Not known

   135 _____   ⬚ mU/L   ⬚ µU/ml

136 Was any testing for autoantibodies performed prior to mobilization for stem cell collection?

  ⬚ yes   ⬚ no   ⬚ Unknown

   Specify the test results for the following autoantibodies:

   137 Anti-centromere:

    ⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

   138 Anti-DNA topoi-
     somerase I (Scl-70):

    ⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

   139 Anti-nuclear:

    ⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

   140 Anti-SS-A:

    ⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

   141 Anti-SS-B:

    ⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

Specify the results of the following pulmonary function tests performed prior to mobilization therapy for stem cell collection:

142 Was dyspnea present on exertion?

  ⬚ yes   ⬚ no   ⬚ Unknown

143 Vital capacity (VC):

  ⬚ Known   ⬚ Not known

   144 _____ % (predicted value)

   145 Was the actual VC value in the normal range
      (≥ 80 % of predicted value)?

    ⬚ yes   ⬚ no

146 D^{L}CO:

  ⬚ Known   ⬚ Not known

   147 _____ % (predicted value)

148 Was the actual D^LCO value in the normal range
(>=80% or predicted value)?

☐ yes   ☐ no

149 D^LCO corrected for hemoglobin

☐ Known   ☐ Not known

150 _____ % (predicted value)

151 Was the D^LCO value (corrected for hemoglobin)
in the normal range (>=80% of predicted value)?

☐ yes   ☐ no

152 Was oxygen desaturation present on excercize testing?

☐ yes   ☐ no   ☐ Unknown

153 Was ground glass appearance present on chest x-ray?

☐ yes   ☐ no   ☐ Unknown

154 Was a high resolution chest CT scan performed?

☐ yes   ☐ no   ☐ Unknown

155 Was ground glass appearance present on CT scan?

☐ yes   ☐ no   ☐ Unknown

156 Was bronchoalveolar lavage (BAL) performed?

☐ yes   ☐ no   ☐ Unknown

157 Was alveolitis present?

☐ yes   ☐ no

158 Specify the highest percentage of neutrophils present in the _____ %
segments tested:

159 Was pulmonary artery hypertension present?

☐ yes   ☐ no   ☐ Unknown

160 Specify the mean pulmonary artery pressure (PAP) level:

☐ Known   ☐ Unknown

161 _____ mm / Hg

162 Specify the method used to examine the PAP level:

☐ echocardiogram   ☐ catheterization

163 Was systemic hypertension present that required treatment?

☐ yes   ☐ no   ☐ Unknown

Specify treatment(s) given for hypertension:

164 ACE inhibitor

☐ yes   ☐ no

165 Other antihypertensive therapy

☐ yes   ☐ no

166 Specify antihypertensive therapy: _____

167 Specify the duration of antihypertensive therapy: _____ months

168 Was arrhythmia present that required treatment?

☐ yes   ☐ no   ☐ Unknown

169 Was an echocardiogram performed prior to mobilization therapy for stem cell collection?

☐ yes   ☐ no   ☐ Unknown

170 Was pericardial effusion present?

☐ yes   ☐ no   ☐ Unknown

171 Specify the size of the area of accumulated excess fluid:

☐ small   ☐ moderate   ☐ large

172 Specify the left ventricular ejection fraction:

☐ Known   ☐ Not known

173 _____ %

174 Was a multiple gate acquisition scan (MUGA test / nuclear ventriculography) performed prior to mobilization therapy for stem cell collection?

☐ yes   ☐ no   ☐ Unknown

175 Specify the left ventricular ejection fraction: _____ %

176 Did the recipient complete a modified Health Assessment Questionnaire (HAQ) for Scleroderma prior to mobilazation therapy for stem cell collection? *Steen VD, Medsger Jr. TA. The value of the Health Assessment Questionnaire and special patient-generated scales to demonstrate change in systemic sclerosis patients over time. Arthritis Rheum 1997; 40 (11): 1984-1991.*

☐ yes   ☐ no   ☐ Unknown

177 Recipient's score: _____

178 Worst possible score: _____

179 Best possible score: _____

Most Recent Disease Assessment Prior to the Start of the Preparative Regimen	Questions: 180 - 212

Information for this section should come from the most recent evaluation performed < 2 weeks prior to the preparative regimen. If the recipient was not evaluated prior to the preparative regimen, check here ☐ and continue with question 268.

180 Date of evaluation prior to the preparative regimen: __ __ __ __ - __ __- __ __

181 What was the extent of cutaneous systemic sclerosis prior to the start of the preparative regimen?

☐ limited (cutaneous thickening distal (but not proximal) to elbows or knees)

☐ diffuse

☐ Unknown

Specify skin thickness for the following sites as determined by clinical palpation performed prior to the start of the preparative regimen: *Clements P, Lachenbruch P, Seibold J, White B, Weiner S, Martin R, et al. Inter and intraobserver variability of total skin thickness score (modified Rodnan TSS) in systemic sclerosis. J Rheumatol 1995; 22:1281-1285.*

182 Face

☐ score 0 - Normal

☐ score 1 - Mild

☐ score 2 - Moderate

☐ score 3 - Severe (inability to pinch skin into a fold)

183 Anterior chest

☐ score 0 - Normal

☐ score 1 - Mild

☐ score 2 - Moderate

☐ score 3 - Severe (inability to pinch skin into a fold)

184 Abdomen

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

185 Upper arm - left

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

186 Upper arm - right

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

187 Forearms - left

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

188 Forearms - right

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

189 Dorsum of hand - left

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

190 Dorsum of hand - right

    score 0 - Normal

    score 1 - Mild

    score 2 - Moderate

    score 3 - Severe (inability to pinch skin into a fold)

191 Fingers - left

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

192 Fingers - right

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

193 Thigh - left

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

194 Thigh - right

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

195 Lower leg - left

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

196 Lower leg - right

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

197 Dorsum of foot - left

  score 0 - Normal

  score 1 - Mild

  score 2 - Moderate

  score 3 - Severe (inability to pinch skin into a fold)

198 Dorsum of foot - right

ꙮ score 0 - Normal

ꙮ score 1 - Mild

ꙮ score 2 - Moderate

ꙮ score 3 - Severe (inability to pinch skin into a fold)

199 Total modified Rodnan Skin Score: (add scores from questions 182-198) _____

Specify the following clinical findings prior to the start of the preparative regimen:

200 Changes in skin pigmentation:

ꙮ Present   ꙮ Absent   ꙮ Unknown

201 Raynaud's phenomenon:

ꙮ Present   ꙮ Absent   ꙮ Unknown

202 Painful digital ulcers:

ꙮ Present   ꙮ Absent   ꙮ Unknown

203 Specify number of digital ulcers: _____

204 Gut dysmotility:

ꙮ Present   ꙮ Absent   ꙮ Unknown

205 Malabsorption:

ꙮ Present   ꙮ Absent   ꙮ Unknown

206 Weight loss > 10% of body weight:

ꙮ Present   ꙮ Absent   ꙮ Unknown

207 Muscle weakness:

ꙮ Present   ꙮ Absent   ꙮ Unknown

208 Joint tenderness:

ꙮ Present   ꙮ Absent   ꙮ Unknown

209 Specify number of joints affected: _____

210 Tendon friction rubs:

ꙮ Present   ꙮ Absent   ꙮ Unknown

211 Specify the number of sites affected: _____

212 Contractures:

ꙮ Present   ꙮ Absent   ꙮ Unknown

Laboratory Studies Prior to the Start of the Preparative Regimen	Questions: 213 - 263

213 Date lab studies were performed: __ __ __ __ - __ __- __ __

214 Creatinine clearance:

ꙮ Known   ꙮ Not known

215 _____ ꙮ mL/min   ꙮ mL/sec

216 Creatinine phosphokinase:

ꙮ Known   ꙮ Not known

217 _____ ꙮ U/L   ꙮ μkat/L

218 Blood urea nitrogen:

⬚ Known   ⬚ Not known

219 _____ ⬚ mg/dL   ⬚ mmol/L

220 Was there evidence of hematuria prior to the start of the preparative regimen?

⬚ yes, present   ⬚ no, absent   ⬚ Unknown

221 Was there evidence of proteinuria prior to the start of the preparative regimen?

⬚ yes, present   ⬚ no, absent   ⬚ Unknown

222 Thyroid stimulating hormone (TSH):

⬚ Known   ⬚ Not known

223 _____ ⬚ mU/L   ⬚ µU/ml

224 Was any testing for autoantibodies performed prior to the start of the preparative regimen?

⬚ yes   ⬚ no   ⬚ Unknown

Specify the test results for the following autoantibodies:

225 Anti-centromere:

⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

226 Anti-DNA topoisomerase I (Scl-70):

⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

227 Anti-nuclear:

⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

228 Anti-SS-A:

⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

229 Anti-SS-B:

⬚ Positive   ⬚ Negative   ⬚ Inconclusive   ⬚ not tested / unknown

Specify the results of the following pulmonary function tests performed prior to the start of the preparative regimen:

230 Was dyspnea present on exertion?

⬚ yes   ⬚ no   ⬚ Unknown

231 Vital capacity (VC):

⬚ Known   ⬚ Not known

232 _____ % (predicted value)

233 Was the actual VC value in the normal range
(>= 80% of predicted value)?

⬚ yes   ⬚ no

234 D^LCO:

⬚ Known   ⬚ Not known

235 _____ % (predicted value)

236 Was the actual D^LCO: value in the normal range
(>= 80% of the predicted value)?

⬚ yes   ⬚ no

237 D^{L}CO corrected for hemoglobin:

☐ Known   ☐ Not known

238 _____ % *(predicted value)*

239 Was the D^{L}CO value (corrected for hemoglobin) in the normal range (>= 80% of predicted value)?

☐ yes   ☐ no

240 Was oxygen desaturation present on exercise testing?

☐ yes   ☐ no   ☐ Unknown

241 Was ground glass appearance present on chest x-ray?

☐ yes   ☐ no   ☐ Unknown

242 Was a high resolution chest CT performed?

☐ yes   ☐ no   ☐ Unknown

243 Was ground glass appearance present on CT scan?

☐ yes   ☐ no   ☐ Unknown

244 Was bronchoalveolar lavage (BAL) performed?

☐ yes   ☐ no   ☐ Unknown

245 Was alveolitis present?

☐ yes   ☐ no

246 Specify the highest percentage of neutrophils present in the segments tested: _____ %

247 Was pulmonary artery hypertension present?

☐ yes   ☐ no   ☐ Unknown

248 Specify the mean pulmonary artery pressure (PAP) level:

☐ Known   ☐ Not known

249 _____ mm / Hg

250 Specify the method used to examine the PAP level:

☐ echocardiogram   ☐ catheterization

251 Was systemic hypertension present that required treatment?

☐ yes   ☐ no   ☐ Unknown

Specify treatment(s) given for hypertension:

252 ACE inhibitor

☐ yes   ☐ no

253 Other antihypertensive therapy

☐ yes   ☐ no

254 Specify antihypertensive therapy: _____

255 Specify the duration of antihypertensive therapy: _____ months

256 Was arrhythmia present that required treatment?

☐ yes   ☐ no   ☐ Unknown

257 Was an echocardiogram performed prior to the start of the preparative regimen?

☐ yes   ☐ no   ☐ Unknown

258 Was pericardial effusion present?

☐ yes   ☐ no   ☐ Unknown

259 Specify the size of the area of accumulated excess fluid:

☐ small   ☐ moderate   ☐ large

260 Specify the left ventricular ejection fraction:

☐ Known   ☐ Not known

261 _____ %

262 Was a multiple gate acquisition scan (MUGA test / nuclear ventriculography) performed prior to the start of the preparative regimen?

☐ yes   ☐ no   ☐ Unknown

263 Sepcify the left ventricular ejection fraction: _____ %

Functional Assessment Prior to the Start of the Preparative Regimen	Questions: 264 - 274

264 Did the recipient complete a modified Health Assessment Questionnaire (HAQ) for Scleroderma prior to the start of the preparative regimen? *Steen VD, Medsger Jr. TA. The value of the Health Assessment Questionnaire and special patient-generated scales to demonstrate change in systemic sclerosis patients over time. Arthritis Rheum 1997; 40 (11): 1984-1991.*

☐ yes   ☐ no   ☐ Unknown

265 Recipient's score: _____

266 Worst possible score: _____

267 Best possible score: _____

Specify the site(s) of disease involvement which were included in the primary indication(s) to proceed with HSCT:

268 Gastrointestinal (GI) tract

☐ yes   ☐ no   ☐ Unknown

269 Heart

☐ yes   ☐ no   ☐ Unknown

270 Lungs

☐ yes   ☐ no   ☐ Unknown

271 Severe functional impairment

☐ yes   ☐ no   ☐ Unknown

272 Skin

☐ yes   ☐ no   ☐ Unknown

273 Other site of disease

☐ yes   ☐ no   ☐ Unknown

274 Specify site of disease: _____

First Name: _____   Last Name: _____

Phone number: _____   Fax number: _____

E-mail address: _____