

Form 2144 R2.0: Systemic Sclerosis Post-HSCT Data

Center:

CRID:

Key Pages

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: _____

Visit:

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

Specify: _____

Disease Assessment Post-HSCT

Questions: 1 - 1

1 Specify the date the recipient was evaluated for this report: ____ - ____ - ____

Post-HSCT Treatment for Systemic Sclerosis

Questions: 2 - 58

2 Did the recipient receive any treatment for systemic sclerosis since the date of the last report?

☐ yes ☐ no ☐ Unknown

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

☐ yes ☐ no ☐ Unknown

4 Reason for therapy: _____

5 Specify other reason: _____

6 Date therapy started: ____ - ____ - ____

7 Currently Receiving?

☐ yes ☐ no

8 Cyclophosphamide (CTX, Cytoxan, Neosar)

☐ yes ☐ no ☐ Unknown

9 Reason for therapy: _____

10 Specify other reason: _____

11 Date therapy started: ____ - ____ - ____

12 Currently Receiving?

☐ yes ☐ no

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13 Cyclosporine (CsA, Neoral, Sandimmune)

☐ yes ☐ no ☐ Unknown

14 Reason for therapy: _____

15 Specify other reason: _____

16 Date therapy started: ____ - ____ - ____

17 Currently Receiving?

☐ yes ☐ no

18 D-penicillamine (penicilamine, Cuprimine, Depen)

☐ yes ☐ no ☐ Unknown

19 Reason for therapy: _____

20 Specify other reason: _____

21 Date therapy started: ____ - ____ - ____

22 Currently Receiving?

☐ yes ☐ no

23 Methotrexate (MTX, Folex)

☐ yes ☐ no ☐ Unknown

24 Reason for therapy: _____

25 Specify other reason: _____

26 Date therapy started: ____ - ____ - ____

27 Currently Receiving?

☐ yes ☐ no

28 Mycophenolate mofetil (MMF, CellCept)

☐ yes ☐ no ☐ Unknown

29 Reason for therapy: _____

30 Specify other reason: _____

31 Date therapy started: ____ - ____ - ____

32 Currently Receiving?

☐ yes ☐ no

33 Phototherapy

☐ yes ☐ no ☐ Unknown

34 Reason for therapy: _____

35 Specify other reason: _____

36 Date therapy started: ____ - ____ - ____

37 Currently Receiving?

☐ yes ☐ no

38 Prednisone (Intensol, Sterapred) or equivalent

☐ yes ☐ no ☐ Unknown

39 Reason for therapy: _____

40 Specify other reason: _____

41 Date therapy started: ____ - ____ - ____

42 Currently Receiving?

☐ yes ☐ no

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43 Prostanoids/prostaglandin analogs

yes no Unknown

44 Reason for therapy:

45 Specify other reason:

46 Date therapy started: - - - - - - - -

47 Currently Receiving?

yes no

48 Tacrolimus (FK 506, Prograf)

yes no Unknown

49 Reason for therapy:

50 Specify other reason:

51 Date therapy started: - - - - - - - -

52 Currently Receiving?

yes no

53 Other treatment

yes no Unknown

54 Specify other treatment:

55 Reason for therapy:

56 Specify other reason:

57 Date therapy started: - - - - - - - -

58 Currently Receiving?

yes no

Disease Status at the Time of Evaluation for This Reporting Period

Questions: 59 - 91

59 Date of evaluation for this reporting period: - - - - - - - -

60 What was the extent of cutaneous systemic sclerosis at the time of evaluation for this report?

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 at the time of evaluation for this report: 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.

Indicate Modified Rodnan Skin Thickness Score (MRSS) for each anatomic area below:





61 Face

score 0 - Normal
score 1 - Mild
score 2 - Moderate
score 3 - Severe (inability to pinch skin into a fold)





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62 Anterior chest

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





63 Abdomen

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





64 Upper arm-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





65 Upper arm-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





66 Forearm-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)

67 Forearm-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





68 Dorsum of hand-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





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69 Dorsum of hand-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





70 Fingers-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





71 Fingers-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





72 Thigh-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)





73 Thigh-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)

74 Lower leg-left

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)

75 Lower leg-right

-  score 0 - Normal
-  score 1 - Mild
-  score 2 - Moderate
-  score 3 - Severe (inability to pinch skin into a fold)

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76 Dorsum of foot-left

- ☐ score 0 - Normal
- ☐ score 1 - Mild
- ☐ score 2 - Moderate
- ☐ score 3 - Severe (inability to pinch skin into a fold)

77 Dorsum of foot-right

- ☐ score 0 - Normal
- ☐ score 1 - Mild
- ☐ score 2 - Moderate
- ☐ score 3 - Severe (inability to pinch skin into a fold)

78 Total modified Rodnan Skin Score:(add scores from questions 61-77) _____

Specify the following clinical findings at the time of evaluation for this report:

79 Changes in skin pigmentation:

- ☐ Present
- ☐ Absent
- ☐ Unknown

80 Raynaud's phenomenon:

- ☐ Present
- ☐ Absent
- ☐ Unknown

81 Painful digital ulcers:

- ☐ Present
- ☐ Absent
- ☐ Unknown

82 Specify number of digital ulcers _____

83 Gut dysmotility:

- ☐ Present
- ☐ Absent
- ☐ Unknown

84 Malabsorption:

- ☐ Present
- ☐ Absent
- ☐ Unknown

85 Weight loss > 10% of body weight:

- ☐ Present
- ☐ Absent
- ☐ Unknown

86 Muscle weakness

- ☐ Present
- ☐ Absent
- ☐ Unknown

87 Joint tenderness:

- ☐ Present
- ☐ Absent
- ☐ Unknown

88 Specify number of joints affected: _____

89 Tendon friction rubs:

- ☐ Present
- ☐ Absent
- ☐ Unknown

90 Specify number of sites affected: _____

91 Contractures:

- ☐ Present
- ☐ Absent
- ☐ Unknown

Laboratory Studies at the Time of Evaluation for This Reporting Period

Questions: 92 - 140

92 Serum creatinine

- ☐ Known
- ☐ Not known

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93 _____ mg/dL mmol/L μmol/L

94 Creatinine clearance:

☐ Known ☐ Not known

95 _____ mL/min mL/sec

96 Creatinine phosphokinase:

☐ Known ☐ Not known

97 _____ U/L μkat/L

98 Blood urea nitrogen

☐ Known ☐ Unknown

99 _____ mg/dL mmol/L

100 Was there evidence of hematuria at the time of evaluation for this report?

☐ yes, present ☐ no, absent ☐ Unknown

101 Was there evidence of proteinuria at the time of evaluation for this report?

☐ yes, present ☐ no, absent ☐ Unknown

102 Thyroid stimulating hormone (TSH):

☐ Known ☐ Unknown

103 _____ mU/L μU/ml

104 Was any testing for autoantibodies performed since the date of the last report?

☐ yes ☐ no ☐ Unknown

Specify the test results for the following autoantibodies:

105 Anti-centromere:

☐ Positive ☐ Negative ☐ Inconclusive ☐ not tested / unknown

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

☐ Positive ☐ Negative ☐ Inconclusive ☐ not tested / unknown

107 Anti-nuclear:

☐ Positive ☐ Negative ☐ Inconclusive ☐ not tested / unknown

108 Anti-SS-A:

☐ Positive ☐ Negative ☐ Inconclusive ☐ not tested / unknown

109 Anti-SS-B:

☐ Positive ☐ Negative ☐ Inconclusive ☐ not tested / unknown

Specify the results of the following pulmonary function tests performed since the date of the last report:

110 Was dyspnea present on exertion?

☐ yes ☐ no ☐ Unknown

111 Vital capacity (VC):

☐ Known ☐ Unknown

112 _____ % (predicted value)

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

☐ yes ☐ no

114 D^LCO:

☐ Known ☐ Not known

115 _____ % (predicted value)

116 Was the actual D^LCO value in the normal
range (≥ 80% of predicted value)?

☐ yes ☐ no

117 D^LCO corrected for hemoglobin:

☐ Known ☐ Unknown

118 _____ % (predicted value)

119 Was the D^LCO value (corrected for hemoglobin)
in the normal range (≥ 80% of predicted value)?

☐ yes ☐ no

120 Was oxygen desaturation present on exercise testing?

☐ yes ☐ no ☐ Unknown

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

☐ yes ☐ no ☐ Unknown

122 Was a high resolution chest CT scan performed?

☐ yes ☐ no ☐ Unknown

123 Was ground glass appearance present on CT scan?

☐ yes ☐ no ☐ Unknown

124 Was pulmonary artery hypertension present?

☐ yes ☐ no ☐ Unknown

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

☐ Known ☐ Unknown

126 _____ mm/Hg

127 Specify the method used to examine the PAP level:

☐ echocardiogram ☐ catheterization

128 Was systemic hypertension present that required treatment?

☐ yes ☐ no ☐ Unknown

Specify treatment(s) given for hypertension:

129 ACE inhibitor

☐ yes ☐ no

130 Other antihypertensive therapy

☐ yes ☐ no

131 Specify antihypertensive therapy: _____

132 Specify the duration of antihypertensive therapy: _____

133 Was arrhythmia present that required treatment?

☐ yes ☐ no ☐ Unknown

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134 Was an echocardiogram performed at the time of evaluation for this report?

yes no Unknown

135 Was pericardial effusion present?

yes no Unknown

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

small moderate large

137 Specify the left ventricular ejection fraction:

Known Not known

138 %

139 Was a multiple gate acquisition scan (MUGA test/nuclear ventriculography) performed at the time of evaluation for this report?

yes no Unknown

140 Specify the left ventricular ejection fraction: %

Functional Assessment at the Time of Evaluation for This Report

Questions: 141 - 144

141 Did the recipient complete a modified Health Assessment Questionnaire (HAQ) for Scleroderma at the time of evaluation for this report?

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

142 Recipient's score:

143 Worst possible score:

144 Best possible score:

First Name: Last Name:

Phone number: Fax number:

E-mail address: