Disease Assessment at the Time of Best Response to HSCT

1. Specify the percent of clinical improvement at the time of best response since HSCT compared to the evaluation just prior to mobilization, according to American College of Rheumatology (ACR) criteria:
   Giannini EH, Ruperto N, Ravelli A, Lovell DJ, Felson DT, Martini A. Preliminary definition of improvement in juvenile arthritis. Arthritis Rheum 1997; 40 (7): 1202–1209. Requires 20%* or more improvement in 3 of following 6 criteria, with no more than 1 of the remaining variables worsened by more than 30%: • physician global assessment of disease activity • parent / recipient global assessment of overall well-being • functional ability • number of joints with active arthritis • number of joints with limited range of motion • erythrocyte sedimentation rate (ESR).

   * Substitute 50% or 70% for 20% and 50% improvement levels, respectively.

   disease is worse
   no improvement
   20% improvement (ACR20)
   50% improvement (ACR50)
   70% improvement (ACR70)
   disease in remission

2. Date of assessment for best response:

3. Specify the date of disease progression:

4. Specify the date of maximal improvement:

5. Specify the date of disease remission:

6. Date of assessment for best response:

To be completed in conjunction with a Form 2100 – 100 Days Post-HSCT Data, Form 2200 – Six Months to Two Years Post-HSCT Data, or Form 2300 – Yearly Follow-Up for Greater Than Two Years Post-HSCT Data. Information reported here should reflect the date of last contact as reported in the post-HSCT data collection form, or immediately prior to death.
### Post-HSCT Treatment for Juvenile Idiopathic Arthritis

6. Did the recipient receive any treatment for JIA / JRA since the date of the last report?

<table>
<thead>
<tr>
<th>Therapy Given?</th>
<th>Reason for Therapy Code</th>
<th>Therapy Code</th>
<th>Date Therapy Started</th>
<th>Currently Receiving?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corticosteroids</td>
<td>yes</td>
<td>Continue with question 43</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>no</td>
<td>Continue with table below</td>
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</tr>
<tr>
<td></td>
<td>unknown</td>
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</tr>
<tr>
<td>Cyclosphosphamide (CTX, Cytoxan, Neosar)</td>
<td>yes</td>
<td></td>
<td>20</td>
<td>11. yes</td>
</tr>
<tr>
<td></td>
<td>no</td>
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</tr>
<tr>
<td></td>
<td>unknown</td>
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</tr>
<tr>
<td>Cyclosporine (CsA, Neoral, Sandimmune)</td>
<td>yes</td>
<td></td>
<td>20</td>
<td>21. yes</td>
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<tr>
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</tr>
<tr>
<td></td>
<td>unknown</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Etanercept (Enbrel)</td>
<td>yes</td>
<td></td>
<td>20</td>
<td>26. yes</td>
</tr>
<tr>
<td></td>
<td>no</td>
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</tr>
<tr>
<td></td>
<td>unknown</td>
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<td></td>
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<tr>
<td>Methotrexate (MTX, Folex)</td>
<td>yes</td>
<td></td>
<td>20</td>
<td>31. yes</td>
</tr>
<tr>
<td></td>
<td>no</td>
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</tr>
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<td></td>
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<tr>
<td>Other treatment</td>
<td>yes</td>
<td></td>
<td>20</td>
<td>41. yes</td>
</tr>
<tr>
<td></td>
<td>no</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>unknown</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

#### Reason for Therapy Codes

1. Planned per protocol  
2. Continued from prior to HSCT  
3. Relapse / progression of JRA  
4. Other reason  
5. Reason unknown
Disease Status at the Time of Assessment for This Reporting Period

43. Date of assessment for the current disease evaluation:  

44. Specify the number of painful / tender joints since the date of the last report:

45. Specify the number of swollen / effused joints since the date of the last report:

46. Specify the Pediatric EPM Range of Motion final score (0.0–3.0):

47. Was morning stiffness present since the date of the last report?

48. Specify the duration of morning stiffness:

Laboratory Studies for This Reporting Period

Specify if any of the following laboratory values were elevated since the date of the last report:

51. 1 yes 2 no 3 unknown C-reactive protein

52. 1 yes 2 no 3 unknown Erythrocyte sedimentation rate (ESR)

Specify the results of the following immune function studies performed since the date of the last report:

Quantitative immunoglobulins:

53. IgG 1 normal 2 decreased 3 increased 4 unknown

54. IgA 1 normal 2 decreased 3 increased 4 unknown

55. IgM 1 normal 2 decreased 3 increased 4 unknown

56. IgE 1 normal 2 decreased 3 increased 4 unknown

Lymphocyte subsets:

57. CD3 1 normal 2 decreased 3 increased 4 unknown

58. CD4 1 normal 2 decreased 3 increased 4 unknown

59. CD8 1 normal 2 decreased 3 increased 4 unknown

60. CD16 1 normal 2 decreased 3 increased 4 unknown

61. CD19 1 normal 2 decreased 3 increased 4 unknown
Radiographic Assessment for This Reporting Period

62. Were radiographic bone erosions present since the date of the last report?
   1. yes
   2. no
   3. unknown

63. Was advanced skeletal age of affected joints noted radiographically since the date of the last report?
   1. yes
   2. no
   3. unknown

64. Was osteoporosis present since the date of the last report?
   1. yes
   2. no
   3. unknown

65. Were osteoporotic fractures present?
   1. yes
   2. no
   3. unknown

Functional Assessment for This Reporting Period

66. Did the recipient complete a Childhood Health Assessment Questionnaire (CHAQ) since the date of the last report?
   1. yes
   2. no
   3. unknown

Specify the following scores for the CHAQ pain sub-scale:

67. Recipient’s pain assessment:
68. Worst possible pain score:
69. Best possible pain score:

Specify the following scores for the CHAQ disability sub-scale:

70. Recipient’s disability assessment:
71. Worst possible disability score:
72. Best possible disability score:

Specify the following scores for the CHAQ severity sub-scale:

73. Recipient’s severity assessment:
74. Worst possible severity score:
75. Best possible severity score:

76. Did the physician complete a Global Assessment of Functioning of the recipient’s health since the date of the last report?
   1. yes
   2. no
   3. unknown

77. Physician-rated Global Assessment score:
78. Worst possible score:
79. Best possible score:
80. Specify the percent of clinical improvement since the date of the last report, according to American College of Rheumatology (ACR) criteria:

Giannini EH, Ruperto N, Ravelli A, Lovell DJ, Felson DT, Martini A. Preliminary definition of improvement in juvenile arthritis. Arthritis Rheum 1997; 40 (7): 1202–1209. Requires 20%* or more improvement in 3 of following 6 criteria, with no more than 1 of the remaining variables worsened by more than 30%*: • physician global assessment of disease activity • parent / recipient global assessment of overall well-being • functional ability • number of joints with active arthritis • number of joints with limited range of motion • erythrocyte sedimentation rate (ESR).

* Substitute 50% or 70% for 50% and 70% improvement levels, respectively.

1. worse
2. no improvement
3. 20% improvement (ACR20)
4. 50% improvement (ACR50)
5. 70% improvement (ACR70)
6. disease in remission

81. Specify the date of disease progression:

82. Specify the date of maximal improvement:

83. Specify the date of disease remission:

84. Signed: ____________________________

Person completing form

Please print name: ______________________

Phone number: (_______) __________________

Fax number: (_______) __________________

E-mail address: ________________________

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).