Disease Assessment at the Time of Best Response to HSCT

1. Compared to the disease status prior to the preparative regimen, what was the best response to HSCT since the date of the last report? (Include response to any post-HSCT treatment planned as of Day 0.)

   1/3 complete response — no primary tumor, no metastatic sites, catecholamines normal; includes continued complete response
   2/3 very good partial response — primary tumor decreased by 90-99%, no metastatic sites, catecholamines normal; residual 99Tc bone changes allowed
   3/3 partial response — primary tumor decreased by > 50%, all measurable metastatic sites decreased by > 50%, number of positive bone sites decreased by > 50%, no more than 1 positive bone marrow site allowed, 1 positive marrow aspirate or biopsy allowed if this represents a decrease from the number of positive sites at diagnosis
   4/3 minimal response — no new lesions; > 50% reduction of any measurable lesion (primary or metastases) with < 50% reduction in any other; < 25% increase in any existing lesion
   5/3 no response — no new lesions; < 50% reduction but < 25% increase in any existing lesion
   6/3 progressive disease — any new lesions; increase of any measurable lesion by > 25%; previous negative marrow positive for tumor
   7/3 not assessed
   8/3 not tested/unknown

Best response is based on response to the HSCT, but does NOT include response to any therapy given for disease relapse or progression post-HSCT. When determining the best response to HSCT, compare the post-HSCT disease status to the status immediately prior to the preparative regimen, regardless of time since HSCT. This comparison is meant to capture the BEST disease status in response to HSCT that occurred in the reporting interval, even if a subsequent disease relapse or progression occurred during the same reporting interval. If a recipient already achieved their best response in a previous reporting interval, confirm the best response and check the box to indicate “date previously reported.”

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.

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.
18. Specify the date best response was determined: Month Day Year  
   □ date previously reported

19. Were tumor markers evaluated for the best response post-HSCT determination?
   1 □ yes  
   2 □ no

<table>
<thead>
<tr>
<th>Tumor Marker Analysis</th>
<th>Known or Not Known</th>
<th>µg/mg or ng/mL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Homovanillic acid (HVA)</td>
<td>1 □ known 2 □ not known</td>
<td>µg/mg creatinine</td>
</tr>
<tr>
<td>Neuron specific enolase</td>
<td>1 □ known 2 □ not known</td>
<td>ng/mL</td>
</tr>
<tr>
<td>Serum ferritin</td>
<td>1 □ known 2 □ not known</td>
<td>ng/mL or µg/L</td>
</tr>
<tr>
<td>Vanilmandelic acid (VMA)</td>
<td>1 □ known 2 □ not known</td>
<td>µg/mg creatinine</td>
</tr>
</tbody>
</table>

20. Specify the following tumor marker analyses performed:  
   Date of best response determination: Month Day Year  

21. Specify other analysis: ____________________________  
   Specify level and units: ____________________________

31. Was the recipient given planned per protocol post-HSCT treatment for neuroblastoma?
   1 □ yes  
   2 □ no

32. Was radiotherapy given?
   1 □ yes  
   2 □ no

<table>
<thead>
<tr>
<th>Radiotherapy Site</th>
<th>Yes or No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone metastases</td>
<td>1 □ yes 2 □ no</td>
</tr>
<tr>
<td>Primary tumor</td>
<td>1 □ yes 2 □ no</td>
</tr>
<tr>
<td>Other site</td>
<td>1 □ yes 2 □ no</td>
</tr>
</tbody>
</table>

33. Specify the site(s) of radiotherapy: ____________________________

34. Specify the date radiotherapy was started: Month Day Year  

35. Number of fractions given: ____________________________  
   Dose per fraction: ____________________________ cGy (rads)

36. Specify: ____________________________

37. Specify the date MIBG treatment was performed: Month Day Year  

40. Was MIBG given?
   1 □ yes  
   2 □ no

<table>
<thead>
<tr>
<th>Radioisotope</th>
<th>Yes or No</th>
</tr>
</thead>
<tbody>
<tr>
<td>131I-MIBG</td>
<td>1 □ yes 2 □ no</td>
</tr>
<tr>
<td>Other</td>
<td>1 □ yes 2 □ no</td>
</tr>
</tbody>
</table>

41. Specify the radioisotope given: ____________________________

42. Specify: ____________________________

43. Specify: ____________________________

44. Specify the date MIBG treatment was performed: Month Day Year  

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
45. Were retinoids given?
   1 [ ] yes
   2 [ ] no

46. Specify the retinoids given:
   1 [ ] yes
   2 [ ] no
   Isotretinoin

47. Specify the drug(s) given:
   1 [ ] yes
   2 [ ] no
   Other

48. Specify:

49. Specify the date retinoid treatment was started:
   Month Day Year

50. Was immunotherapy given?
   1 [ ] yes
   2 [ ] no

51. Specify the drug(s) given:
   1 [ ] yes
   2 [ ] no
   α-interferon

52. Specify the drug(s) given:
   1 [ ] yes
   2 [ ] no
   Anti-GD2 antibody CH14.18

53. Specify the drug(s) given:
   1 [ ] yes
   2 [ ] no
   Interleukin-2 (IL-2)

54. Specify the drug(s) given:
   1 [ ] yes
   2 [ ] no
   Other

55. Specify:

56. Specify the date immunotherapy was started:
   Month Day Year

57. Was chemotherapy given?
   1 [ ] yes
   2 [ ] no

58. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Adriamycin

59. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Cisplatin

60. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Cyclophosphamide

61. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Dacarbazine (DTIC)

62. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Etoposide (VP16)

63. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Ifosfamide

64. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Melphalan (L-PAM)

65. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Teniposide (VM26)

66. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Vinca alkaloids

67. Specify the treatment(s) given:
   1 [ ] yes
   2 [ ] no
   Other

68. Specify:

69. Specify the date chemotherapy was started:
   Month Day Year

70. Was other treatment given?
   1 [ ] yes
   2 [ ] no

71. Specify other treatment:

72. Specify the date other treatment was started:
   Month Day Year

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
73. Did the neuroblastoma recur or progress since the date of the last report?

1 [ ] yes 2 [ ] no

<table>
<thead>
<tr>
<th>Specify the known site(s) of disease progression / recurrence:</th>
<th>Date determined:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Month</td>
<td>Day</td>
</tr>
<tr>
<td>74. 1 [ ] yes 2 [ ] no Adrenal gland</td>
<td>75.</td>
</tr>
<tr>
<td>76. 1 [ ] yes 2 [ ] no Bone</td>
<td>77.</td>
</tr>
<tr>
<td>78. 1 [ ] yes 2 [ ] no Bone marrow</td>
<td>79.</td>
</tr>
<tr>
<td>80. 1 [ ] yes 2 [ ] no Cerebellum</td>
<td>81.</td>
</tr>
<tr>
<td>82. 1 [ ] yes 2 [ ] no Cerebrospinal fluid (CSF)</td>
<td>83.</td>
</tr>
<tr>
<td>84. 1 [ ] yes 2 [ ] no Cerebrum</td>
<td>85.</td>
</tr>
<tr>
<td>86. 1 [ ] yes 2 [ ] no Cranial nerves</td>
<td>87.</td>
</tr>
<tr>
<td>88. 1 [ ] yes 2 [ ] no Liver</td>
<td>89.</td>
</tr>
<tr>
<td>90. 1 [ ] yes 2 [ ] no Lymph nodes</td>
<td>91.</td>
</tr>
<tr>
<td>92. 1 [ ] yes 2 [ ] no Mediastinum</td>
<td>93.</td>
</tr>
<tr>
<td>94. 1 [ ] yes 2 [ ] no Paraspinal ganglion</td>
<td>95.</td>
</tr>
<tr>
<td>96. 1 [ ] yes 2 [ ] no Retro-orbital area</td>
<td>97.</td>
</tr>
<tr>
<td>98. 1 [ ] yes 2 [ ] no Skin / subcutaneous tissue</td>
<td>99.</td>
</tr>
<tr>
<td>100. 1 [ ] yes 2 [ ] no Other site</td>
<td>101.</td>
</tr>
</tbody>
</table>

102. Specify other site: __________________________

Specify the methods used to examine sites of disease recurrence / persistence / progression: Specify disease status:

103. 1 [ ] yes 2 [ ] no Biopsy | 104. 1 [ ] positive 2 [ ] negative |
| 105. 1 [ ] yes 2 [ ] no Bone scan | 106. 1 [ ] positive 2 [ ] negative |
| 107. 1 [ ] yes 2 [ ] no Radiology | 108. 1 [ ] positive 2 [ ] negative |
| 109. 1 [ ] yes 2 [ ] no Other method | 110. 1 [ ] positive 2 [ ] negative |

111. Specify other method: __________________________
### 112. Was the recipient given treatment for post-HSCT persistent, progressive or recurrent disease since the date of the last report?

- 1: yes
- 2: no
- 3: unknown

### 113. Was radiotherapy given?

- 1: yes
- 2: no

Specify the site(s) of radiotherapy:
- 114. Bone metastases
- 115. Primary tumor
- 116. Other

Specify the date radiotherapy was started:

Specify the dose per fraction:

Specify the radioisotope given:
- 122. 131I-MIBG
- 123. Other

Specify the date MBIG treatment was performed:

### 121. Was MIBG given?

- 1: yes
- 2: no

Specify the date MIBG treatment was performed:

### 123. Were retinoids given?

- 1: yes
- 2: no

Specify the retinoids given:
- Isotretinoin
- Other

Specify the date retinoid treatment was started:

### 131. Was immunotherapy given?

- 1: yes
- 2: no

Specify the drug(s) given:
- α-interferon
- Anti-GD2 antibody CH14.18
- Interleukin-2 (IL-2)
- Other

Specify the date immunotherapy was started:

### 138. Was chemotherapy given?

- 1: yes
- 2: no

Specify the treatment(s) given:
- Adriamycin
- Cisplatin
- Cyclophosphamide
- Dacarbazine (DTIC)
- Etoposide (VP16)
- Ifosfamide
- Melphalan (L-PAM)
- Teniposide (VM26)
- Vincristine

Specify the date chemotherapy was started:
154. What is the current disease status?
   1 □ complete remission
   2 □ not in complete remission

155. Date the current disease status was established in this reporting period:  
   [ ] [ ] [ ] Month  [ ] [ ] 20 [ ] Day  [ ] [ ] Year

156. Signed: ______________________________________________________  

   Person completing form

   Please print name: _________________________________________________

   Phone: (_________ ) _____________________________________________

   Fax: (_________ ) _______________________________________________

   E-mail address: ___________________________________________________