Leukodystrophies
Pre-HSCT Data

1. What was the date of diagnosis of Leukodystrophy?

2. Specify the leukodystrophy subtype:
   1. globoid cell leukodystrophy
   2. metachromatic leukodystrophy

   Specify the leukocyte galactocerebrosidase enzyme activity at diagnosis:
   3. Date recipient tested:
      1. known
      2. not known
   4. Recipient result:
      1. known
      2. not known
   5. Donor result:
      1. known
      2. not known

   Specify the leukocyte arylsulfatase A enzyme activity at diagnosis:
   6. Date recipient tested:
      1. known
      2. not known
   7. Recipient result:
      1. known
      2. not known
   8. Were the recipient's urinary sulfatides elevated at diagnosis?
      1. yes
      2. no
      3. not known

This form must be accompanied by Form 2000 – Recipient Baseline Data. All information in the box above, including the date, should be identical with the corresponding Form 2000. Information should come from an actual examination by the Transplant Center physician, or the physician who is following the recipient pre-HSCT, or abstraction of the recipient's medical records.

If this is a report of a second or subsequent transplant, check here ☐ and continue with question 24.
### Clinical Status Prior to the Preparative Regimen

24. Is there a history of seizures at any time prior to the preparative regimen?
- **1** yes
- **2** no
- **3** unknown

25. Was cerebrospinal fluid (CSF) testing done prior to the preparative regimen?
- **1** yes
- **2** no
- **3** unknown

#### Specify results of most recent tests:

26. Date of most recent testing:

27. Opening pressure:
- **1** known
- **2** not known

---

9. **Donor** result:
- **1** known
- **2** not known

10. Mean fasting plasma very-long-chain fatty acid (VLCFA) C26:0 level at diagnosis:
- **1** known
- **2** not known

11. Was the acid level measured within two weeks prior to the preparative regimen?
- **1** yes
- **2** no
- **3** unknown

12. Date recipient tested:

13. Plasma level:

14. Was treatment given for adrenal insufficiency between diagnosis and HSCT?
- **1** yes
- **2** no
- **3** unknown

15. Specify adrenal insufficiency:
- **1** yes
- **2** no
- **3** unknown

16. Glucocorticoid

17. Was treatment given to lower plasma very-long-chain fatty acids at any time prior to HSCT?
- **1** yes
- **2** no
- **3** unknown

18. Specify treatment(s):
- **1** yes
- **2** no
- **3** unknown

19. 4-phenylbutyrate

20. GTE:GTO oil (Lorenzo’s oil)

21. Lovastatin or related compound

22. Other

23. **Donor’s** mean fasting VLCFA C26:0 level:
- **1** known
- **2** not known

---

CIBMTR Form 2037 (LDS) v1.0 (2–5) July 2007
Copyright © 2007 National Marrow Donor Program and
The Medical College of Wisconsin, Inc. All rights reserved.
For internal use only. Document P00546 version 1.0 Replaces: n/a
28. Closing pressure:
   1 ☐ known cm H₂O
   2 ☐ not known

29. Total protein:
   1 ☐ known mg/dL
   2 ☐ not known

30. Was Magnetic Resonance Imaging (MRI) performed at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

31. Date of most recent test prior to the preparative regimen:
   Month Day Year

32. Specify MRI results:
   1 ☐ normal
   2 ☐ abnormal
   3 ☐ not known

33. Is a copy of the MRI report attached?
   1 ☐ yes
   2 ☐ no

34. Was Magnetic Resonance Spectroscopy performed at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

35. Date of most recent test prior to the preparative regimen:
   Month Day Year

36. Specify MRS results:
   1 ☐ normal
   2 ☐ abnormal
   3 ☐ not known

37. Is a copy of the MRS report attached?
   1 ☐ yes
   2 ☐ no

38. Were nerve conduction velocities tested at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

39. Date of most recent test prior to the preparative regimen:
   Month Day Year

40. Specify results:
   1 ☐ normal
   2 ☐ abnormal / impaired
   3 ☐ not known

41. Is a copy of the report attached?
   1 ☐ yes
   2 ☐ no
42. Was a Mental Development test administered at any time prior to the preparative regimen?
   1. yes
   2. no
   3. unknown

43. Date of most recent test prior to the preparative regimen:
   Month Day Year

44. Specify test instrument administered:
   1. Bayley Scales of Infant Development
   2. Stanford Binet Intelligence Scale
   3. Wechsler Preschool and Primary Scale of Intelligence (WPPSI – Revised)
   4. Wechsler Intelligence Scale for Children – III (WISC – III)
   5. other test

45. Specify other test:

46. Full scale score: (not percentile)
   1. known
   2. not known

47. Performance score: (not percentile)
   1. known
   2. not known

48. Verbal score: (not percentile)
   1. known
   2. not known

49. Were the Vineland Adaptive Behavior Scales administered at any time prior to the preparative regimen?
   1. yes
   2. no
   3. unknown

50. Date of most recent test prior to the preparative regimen:
   Month Day Year

51. Communication skills:
   1. known
   2. not known

52. Daily living skills
   1. known
   2. not known

53. Socialization skills
   1. known
   2. not known

54. Was visual acuity tested at any time prior to the preparative regimen?
   1. yes
   2. no
   3. unknown

55. Is the recipient blind?
   1. yes
   2. no

56. Date of most recent test prior to the preparative regimen:
   Month Day Year

57. Visual acuity of right eye (OD): (uncorrected vision)
   1. known
   2. not known

58. Visual acuity of left eye (OS): (uncorrected vision)
   1. known
   2. not known

59. Visual acuity of both eyes (OU): (uncorrected vision)
   1. known
   2. not known
60. Did the recipient undergo an ophthalmologic exam under anesthesia at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

61. Date of most recent test prior to the preparative regimen:
   Month   Day   Year

62. Specify results:
   1 ☐ normal
   2 ☐ abnormal / impaired
   3 ☐ not known

63. Is a copy of the report attached?
   1 ☐ yes
   2 ☐ no

64. Was the hearing loss (HL) in decibels (dB) assessed at the speech threshold for 500 hertz (HZ) at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

65. Date of most recent evaluation:
   Month   Day   Year

   Specify tympanometry results: (See Degree of Hearing Loss chart below for scale ranges.)
   66. 1 ☐ normal / mild  2 ☐ moderate / moderately severe  3 ☐ severe / profound  Right ear
   67. 1 ☐ normal / mild  2 ☐ moderate / moderately severe  3 ☐ severe / profound  Left ear

68. Was the hearing loss (HL) in decibels (dB) assessed at the speech threshold for 2000 hertz (HZ) at any time prior to the preparative regimen?
   1 ☐ yes
   2 ☐ no
   3 ☐ unknown

69. Date of most recent evaluation:
   Month   Day   Year

   Specify tympanometry results: (See Degree of Hearing Loss chart below for scale ranges.)
   70. 1 ☐ normal / mild  2 ☐ moderate / moderately severe  3 ☐ severe / profound  Right ear
   71. 1 ☐ normal / mild  2 ☐ moderate / moderately severe  3 ☐ severe / profound  Left ear

72. Signed: ____________________________  Person completing form

   Please print name: ____________________________

   Phone: (___________) ____________________________

   Fax: (___________) ____________________________

   E-mail address: ____________________________