

INSERT XV
Osteopetrosis

FOR REGISTRY USE ONLY:
I.D. - -
Date received: _____

TEAM:
IUBMID:
(Institutional Unique Blood or Marrow Transplant Identification Number)

Registry (circle one): **IBMTR** ABMTR EBMT

Date of transplant for which this form is being completed:
Month Day Year

Date of report:
Month Day Year

Pretransplant Information*

* If this is a report of a second (or subsequent) transplant, check here and go to Q.53

1. Date of diagnosis of primary disease:
Month Day Year

2. Specify the inheritance of osteopetrosis (check one)
- 1 Autosomal recessive
 - 2 Autosomal recessive with carbanhydrase II deficiency
 - 3 Autosomal dominant (affected parent)
 - 8 Unknown

Clinical Findings Any Time Prior to Conditioning

Yes No Unknown

- 3. 1 0 8 Aplastic Anemia
- 4. 1 0 8 Blindness/Visual impairment
- 5. 1 0 8 Convulsions
- 6. 1 0 8 Dentition problems
- 7. 1 0 8 Exophthalmos
- 8. 1 0 8 Fractures
- 9. 1 0 8 Frontal bossing/prominent forehead
- 10. 1 0 8 Gross motor delay
- 11. 1 0 8 Hearing impairment
- 12. 1 0 8 Height below 5th percentile
- 13. 1 0 8 Hepatomegaly
- 14. 1 0 8 Hypertelorism
- 15. 1 0 8 Mental development delay
- 16. 1 0 8 Nasal congestion
- 17. 1 0 8 Osteomyelitis
- 18. 1 0 8 Septicemia
- 19. 1 0 8 Skull circumference above 95th percentile
- 20. 1 0 8 Splenomegaly
- 21. 1 0 8 Strabismus/Nystagmus
- 22. 1 0 8 Other hematologic impairment(s), if yes specify: _____
- 23. 1 0 8 Other, if yes specify: _____

TEAM:

IUBMID:

Radiologic Findings Any Time Prior to Conditioning

Yes No Unknown

- 24. 1 0 8 "Batman sign"/"Sign du masque"
- 25. 1 0 8 Bone-in-bone
- 26. 1 0 8 Cerebral atrophy (by MRI or CT)
- 27. 1 0 8 Craniosynostosis
- 28. 1 0 8 Hydrocephalus
- 29. 1 0 8 Increased general skeletal sclerosis
- 30. 1 0 8 Metaphyseal widening
- 31. 1 0 8 Other, if yes specify: _____

Pretransplant Therapy Any Time Prior to Conditioning

Yes No Unknown

- 32. 1 0 8 Calcitrol
- 34. 1 0 8 Corticosteroids
- 36. 1 0 8 IFN-gamma
- 38. 1 0 8 Other, if yes specify: _____

Duration (months)

Unknown

Date Stopped

month day year

Unknown

- 33.

- 35.

- 37.

- 39.

40. Did patient receive red blood transfusions at any time prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

41. Number (best estimate) of donor exposures:
1 1-5 2 6-10 3 11-20 4 >20 8 Unknown

42. Did patient receive platelet transfusions at any time prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

43. Number (best estimate) of donor exposures:
1 1-5 2 6-10 3 11-20 4 >20 8 Unknown

44. Was a bone marrow biopsy done at any time prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

45. Date of biopsy:

 Unknown
Month Day Year

46. Is bone marrow biopsy normal?
1 Yes 0 No 8 Unknown

Hematologic Findings Immediately Prior to Conditioning

Specify Units

Transfused Unk

- 47. Absolute lymphocyte count:

 1 x10⁹/L (x10³/mm³) 2 x10⁶/L 8
- 48. Absolute neutrophil count (ANC):

 1 x10⁹/L (x10³/mm³) 2 x10⁶/L 8
- 49. Absolute monocyte count:

 1 x10⁹/L (x10³/mm³) 2 x10⁶/L 8
- 50. Reticulocytes:

 % 8

51. Was a bone biopsy performed within 2 weeks prior to conditioning?

- 1 Yes
- 0 No
- 8 Unknown

52. Specify osteoclast numbers:
1 Few/None 2 Normal 3 Increased 8 Unknown

TEAM:

IUBMID:

Posttransplant Evaluation*

** To be completed at 100 days posttransplant or at time of death if death occurs <100 days after transplant*

Status of Disease Resolution

53. Highest serum calcium value posttransplant: . 1 mg/dL 2 mmol/L 3 mEq/L 8 Unk

54. Date serum calcium tested: Unknown
Month Day Year

55. Did hypercalcemia create a clinical problem which made intervention necessary at any time after start of conditioning?
1 Yes
0 No
7 Not applicable
8 Unknown

56. Is skeletal X-ray normal?

1 Yes
0 No
8 Unknown

57. Date of first normal X-ray: Unknown
Month Day Year

58. Compared to last pretransplant X-ray:
1 Improved 2 No change 3 Worse 8 Unknown

59. Date of first X-ray showing evidence of current status: Unknown
Month Day Year

60. Was a bone marrow biopsy done posttransplant?

1 Yes
0 No
8 Unknown

61. Date of biopsy: Unknown
Month Day Year

62. Is bone marrow biopsy normal?
1 Yes 0 No 8 Unknown

63. Indicate change in bone marrow biopsy compared to last biopsy:
1 Improved 2 No change 3 Worse 8 Unknown

64. Did spleen size normalize?

1 Yes
0 No
7 N/A, no splenomegaly pretransplant
8 Unknown

65. Did growth rate improve?

1 Yes
0 No
7 N/A, no growth delay pretransplant
8 Unknown

If Pre- and/or Posttransplant Growth Curves are available, check here , attach copy and reference Q.65

TEAM:

IUBMID:

Status of Disease Resolution (continued)

66. Was a bone biopsy performed posttransplant?

- 1 Yes
- 0 No
- 8 Unknown

67. Specify osteoclast numbers:

- 1 Few/None
- 2 Normal
- 3 Increased
- 8 Unknown

Indicate Change in Clinical Findings Posttransplant

	<u>Improved</u>	<u>No Change</u>	<u>Worse</u>	<u>1st Occurred Post-Tx</u>	<u>Not Evident Pre- & Post-Tx</u>	<u>Unknown</u>	
68.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Aplastic Anemia
69.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Blindness/Visual impairment
70.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Convulsions
71.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Dentition problems
72.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Exophthalmos
73.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Fractures
74.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Frontal bossing/Prominent forehead
75.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Gross motor delay
76.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Hearing impairment
77.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Height at below 5th percentile
78.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Hepatomegaly
79.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Hypertelorism
80.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Mental development delay
81.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Nasal congestion
82.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Osteomyelitis
83.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Septicemia
84.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Skull circumference above 95th percentile
85.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Splenomegaly
86.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Strabismus/Nystagmus
87.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Other hematologic impairment(s), if yes specify: _____
88.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Other, if yes specify: _____

Indicate Change in Radiologic Findings Posttransplant

	<u>Improved</u>	<u>No Change</u>	<u>Worse</u>	<u>1st Occurred Post-Tx</u>	<u>Not Evident Pre- & Post-Tx</u>	<u>Unknown</u>	
89.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	"Batman sign"/"Sign du masque"
90.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Bone-in-bone
91.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Cerebral atrophy (by MRI or CT)
92.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Craniosynostosis
93.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Hydrocephalus
94.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Increased general skeletal sclerosis
95.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Metaphyseal widening
96.	1 <input type="checkbox"/>	2 <input type="checkbox"/>	3 <input type="checkbox"/>	4 <input type="checkbox"/>	7 <input type="checkbox"/>	8 <input type="checkbox"/>	Other, if yes specify: _____