INSERT XII
Immune Deficiency

FOR REGISTRY USE ONLY:
I.D. ☐ – ☐☐☐☐ – ☐☐☐☐☐☐
Date received:

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

Registry (circle one): IBMTR ABMTR

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.4***

1. Date of diagnosis of primary disease: ☐☐ ☐☐ ☐☐☐☐ (Month Day Year)

2. Specify the immune deficiency phenotype:

1 ☐ SCID, ADA deficiency *(<5% ADA enzyme activity in red blood cells [patient has not been transfused], or in white blood cells)*

2 ☐ SCID, Absence of T-cells with normal B-cells *(This phenotype is X-linked. T-cell number in the absence of maternal engraftment <100/mm^3, B-cells are sIg+ and normal in absolute number for age* ***~ or ~*** *defined mutation in IL-2R gamma chain gene)*

3 ☐ SCID, Absence of T- and B-cells *(Total absolute lymphocyte count <500/mm^3, frequently <200/mm^3, T- or B-cells <100/mm^3. Patients with very low numbers of T- and B-cells but a greater proportion of NK cells should be listed as "other".)*

7 ☐ Other, specify *(include Omenn's syndrome, ZAP 70 deficiency, IL2 deficiency, CD7 deficiency, SCID with NK cells, and others)*:

3. Specify the inheritance of immune deficiency:

1 ☐ X-linked *(family history is positive or patient has been documented to have a defect of an X-linked gene causing SCID)*

2 ☐ Autosomal recessive *(all females, males in families with affected females)*

3 ☐ Acquired *(non-inherited)*

8 ☐ Unknown

Hematologic Findings Immediately Prior to Transplant (or Prior to Conditioning, if Used)

		Specify Units		Unknown
4.	WBC: ☐☐☐☐☐☐.☐	1☐ x 10^9/L (x 10^3/mm^3)	2☐ x 10^6/L	☐
5.	Lymphocytes: ☐☐	%		☐
6.	T-cells (CD3 or equivalent): ☐☐	% of Lymphocytes		☐
7.	CD4+ cells: ☐☐	% of Lymphocytes		☐
8.	CD8+ cells: ☐☐	% of Lymphocytes		☐
9.	B-cells (CD19+, CD20+, sIg+): ☐☐	% of Lymphocytes		☐
10.	NK cells (CD16+ or equivalent): ☐☐	% of Lymphocytes		☐

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Immunologic Findings Immediately Prior to Transplant (or Prior to Conditioning, if Used)

11. Specify the mitogen proliferation response *(response of lymphocytes to phytohemagglutinin, concavalin A or pokeweed mitogen)*:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased (11-60% normal) 3 ❑ Normal 8 ❑ Not tested or Unknown

12. Natural killer cell function *(specific cytolysis of NK-sensitive target cells, e.g. K562)*:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased (11-50% normal) 3 ❑ Normal 8 ❑ Not tested or Unknown

13. IgG:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

14. IgM:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

15. IgA:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

16. IgE:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

17. Specific antibody response to any antigen against which recipient has been adequately immunized (e.g. <3 DPT):
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

Clinical Status of Recipient Immediately Prior to Transplant (or Prior to Conditioning, if Used)

18. Was maternal engraftment present?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

19. Was graft vs host disease (GVHD) present?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

> **20.** Was GVHD caused by *(check only one)*:
> 1 ❑ Maternal cells ❑ Unirradiated blood transfusion 8 ❑ Unknown source

21. Did the patient have failure to thrive *(decrease of 0.5 standard deviation in weight on standard growth curve or weight <5th percentile for age)*?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

22. Did the patient experience chronic (protracted) diarrhea *(>6 weeks in duration)*?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

23. Did the patient have respiratory impairment *(need for chronic or intermittent support with O_2 or artificial ventilation and/or presence of persistent interstitial, nodular or lobar pneumonia)*?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

24. Did B-cell lymphoproliferative disorder (BLPD) develop pretransplant?
1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

> **25.** Was BLPD associated with EBV?
> 1 ❑ Yes 0 ❑ No 8 ❑ Unknown (not tested)

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Posttransplant Information*

To be completed 100 days posttransplant, or at time of death if death occurred <100 days posttransplant, or immediately prior to start of high-dose therapy (conditioning) for second transplant if second transplant done <100 days after first transplant.

Status of Hematologic Engraftment

(refers to quantitative analyses utilizing discriminating DNA markers. Peripheral blood cells must undergo separation or sorting into T, B or lymphoid vs myeloid populations to perform this determination. If RFLP analyses indicate only donor type hematopoiesis mark T-cell, B-cell and myeloid as "predominantly or completely donor")

26. Status of T-cell engraftment:
1 ❑ Predominantly or completely donor (≥80% donor chimerism) 2 ❑ Mixed chimerism (5-80% donor)
3 ❑ Only host T-cells detected 8 ❑ Unknown

27. Status of B-cell engraftment:
1 ❑ Predominantly or completely donor (≥80% donor chimerism) 2 ❑ Mixed chimerism (5-80% donor)
3 ❑ Only host B-cells detected 8 ❑ Unknown

28. Status of myeloid engraftment:
1 ❑ Completely donor (≥80% donor chimerism) 2 ❑ Mixed chimerism
3 ❑ Host only 8 ❑ Unknown

Status of Immunologic Reconstitution

29. Specify status of T-cell function *(refers to mitogen proliferation, T-cell cytotoxicity, and/or DTH responses)*:
1 ❑ Absent (≤10% normal responses) 2 ❑ Normal 3 ❑ Partial 8 ❑ Unknown

30. Specify status of B-cell function *(refers to immunoglobulin synthesis, and/or specific antibody production)*:
1 ❑ Absent (≤10% normal responses) 2 ❑ Normal 3 ❑ Partial 8 ❑ Unknown

31. Specify the mitogen proliferation response *(response of lymphocytes to phytohemagglutinin, concavalin A or pokeweed mitogen)*:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased (11-60% normal) 3 ❑ Normal 8 ❑ Not tested or Unknown

32. Natural killer cell function *(specific cytolysis of NK-sensitive target cells, e.g. K562)*:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased (11-50% normal) 3 ❑ Normal 8 ❑ Not tested or Unknown

33. IgG:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

34. IgM:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

35. IgA:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

36. IgE:
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown

37. Specific antibody response to any antigen against which recipient has been adequately immunized (e.g. <3 DPT):
1 ❑ Absent (≤10% normal) 2 ❑ Decreased 3 ❑ Normal 4 ❑ Increased 8 ❑ Not tested or Unknown