1. What was the date of diagnosis of Immune Deficiency (ID)?
2. What is the immune deficiency molecular abnormality?
   1. common gamma chain (γC; CD132) deficiency
   2. adenosine deaminase (ADA) deficiency
   3. Janus kinase 3 (JAK3) deficiency
   4. recombination-activating gene 1 (RAG1) deficiency
   5. recombination-activating gene 2 (RAG2) deficiency
   6. IL-7Rα deficiency
   7. DNA cross-link repair 1C (DCLRE1C) / Artemis deficiency
   8. CD3γ (gamma) deficiency
   9. CD3δ (delta) deficiency
   10. CD3ε (epsilon) deficiency
   11. CD3ζ (zeta)-chain deficiency
   12. zeta-chain (TCR) associated protein kinase 70 kDa (ZAP-70) deficiency
   13. CD25 deficiency
   14. CD45 deficiency
   15. purine nucleoside phosphorylase (PNP) deficiency
   16. Cernunos-XLF / NHEJ1 deficiency
   17. DNA ligase 4 deficiency
   18. DNA-protein kinase catalytic subunit (DNA-PKcs) deficiency
   19. adenylate kinase 2 (AK2) deficiency (reticular dysgenesis)
   20. Omenn syndrome
   21. bare lymphocyte syndrome (MHC class II) deficiency
   22. cartilage-hair hypoplasia (CHH) / metaphyseal dysplasia, McKusick type
   23. Orai1 deficiency
   24. other molecular abnormality
   25. unknown
3. Specify molecular abnormality:
4. Specify other abnormality:
5. Is the mutated protein or enzyme expressed?
   1  yes
   2  no
   3  unknown

6. What is the pattern of inheritance for the genetic disorder?
   1. sporadic (no family history)
   2. x-linked, documented
   3. autosomal recessive, documented
   4. unknown

7. Are the parents of the patient consanguineous (related by blood ancestry)?
   1  yes
   2  no
   3  unknown

8. Are there other blood relatives in the patient’s family with immunodeficiency disease?
   1  yes
   2  no
   3  unknown

Laboratory Studies at Diagnosis
Report findings prior to any first treatment of the primary disease for which the HSCT is being performed.

9. Date CBC tested: 20 (testing done within 6 weeks of diagnosis)
   Month  Day  Year

Specify units:

10. WBC: 1 x 10^9/L (x 10^3/mm^3) 2 x 10^6/L
    not tested

11. Lymphocytes: %
    not tested

12. Eosinophils: %
    not tested

13. Polymorphonuclear leukocytes (PMN): %
    not tested

14. Hemoglobin: 1 g/dL 2 g/L 3 mmol/L
    not tested  transfused RBC < 30 days from date of test

15. Platelets: 1 x 10^9/L (x 10^3/mm^3) 2 x 10^6/L
    not tested  transfused platelets < 7 days from date of test
Immunoglobulin Analysis
Specify the following quantitative immunoglobulins measured prior to any disease treatment:

16. IgG: o not tested
18. IgM: o not tested
20. IgA: o not tested
22. IgE: IU/mL o not tested

24. Did the recipient receive supplemental intravenous immunoglobulins (IVIG) prior to any first treatment of ID?
1 o yes
2 o no
3 o unknown

Lymphocyte Analysis
Specify the following lymphocyte analyses performed prior to any disease treatment:

26. Were lymphocyte analyses performed?
1 o yes
2 o no

27. Date of most recent testing performed:

28. Absolute lymphocyte count:

% of total lymphocytes:

29. CD3 (T cells): – or –
30. CD4 (T helper cells):
31. CD8 (cytotoxic T cells):
32. CD20 (B lymphocyte cells):
33. CD56 (natural killer (NK) cells):
34. CD4+ / CD45RA+ (naive T cells):
35. CD4+ / CD45RO+ (memory T cells):

25. Was therapy ongoing within one month of immunoglobulin testing?
1 o yes
2 o no

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

36. Date antibody responses were assessed: [ ] [ ] [ ]

Absent Low Normal Not tested

1 2 3 4 37. Bacteriophage phi X-174 or other neoantigen
1 2 3 4 38. Diptheria
1 2 3 4 39. Isohemagglutinin anti-A
1 2 3 4 40. Isohemagglutinin anti-B
1 2 3 4 41. Protein conjugated Hib or pneumococcal vaccine
1 2 3 4 42. Tetanus

43. Unconjugated pneumococcal polysaccharide: [ ] / [ ]

Number of serotypes producing a protective level / Total serotypes tested from vaccine

**Lymphocyte Function**

44. Date lymphocyte function was assessed: [ ] [ ] [ ]

Absent Low Normal Not tested

(< 10% of control) (10-30% of control) (> 30% of control) Not tested

1 2 3 4 45. Anti-CD3
1 2 3 4 46. Candida antigen
1 2 3 4 47. Concavalin A (ConA)
1 2 3 4 48. Phytohemagglutinin (PHA)
1 2 3 4 49. Pokeweed mitogen (PWM)
1 2 3 4 50. Tetanus antigen

**Clinical Features Assessed between Diagnosis and the Start of the Preparative Regimen**

Specify the presence of all clinically significant infections identified between diagnosis and the start of the preparative regimen. If any given infection was identified, use the Codes for Commonly Reported Organisms on the following page to report the organism present. Only report an organism once, even if it was identified at the same site in subsequent infections.

For questions 75–87, also report any fungal infections in the Form 2000 – Recipient Baseline Data beginning at question 163.

Copy this chart to report more than three different infections identified at any one site; check here [ ] if additional pages are attached.

**Site of infection?**

51. 1 [ ] yes 2 [ ] no Hepatitis

First organism Second organism Third organism Specify other organism

52. 53. 54. 55. 

56. If hepatitis was present, was it a prominent feature of ID?

1 [ ] yes 2 [ ] no

57. 1 [ ] yes 2 [ ] no Meningitis / encephalitis

58. 59. 60. 61. 

62. If meningitis / encephalitis was present, was it a prominent feature of ID?

1 [ ] yes 2 [ ] no

63. 1 [ ] yes 2 [ ] no Pneumonia

64. 65. 66. 67. 

68. If pneumonia was present, was it a prominent feature of ID?

1 [ ] yes 2 [ ] no
**Clinical Status between Diagnosis and the Preparative Regimen**

88. Did the recipient experience any of the following clinical features (between diagnosis and prior to the preparative regimen)?

<table>
<thead>
<tr>
<th>Feature present?</th>
<th>If present, is the feature prominent?</th>
</tr>
</thead>
</table>
| 89. yes | no
| 90. yes | no
| 91. yes | no
| 92. yes | no
| 93. yes | no
| 94. yes | no
| 95. yes | no
| 96. yes | no
| 97. yes | no
| 98. yes | no
| 99. yes | no
| 100. yes | no
| 101. yes | no
| 102. yes | no
| 103. yes | no
| 104. yes | no
| 105. yes | no
| 106. yes | no
| 107. yes | no
| 108. yes | no
| 109. yes | no
| 110. yes | no
| 111. yes | no
| 112. yes | no
| 113. yes | no
| 114. yes | no
| 115. yes | no
| 116. yes | no
| 117. yes | no
| 118. yes | no
| 119. yes | no
| 119. yes | no
| 120. yes | no
| 121. yes | no
| 122. yes | no
| 123. yes | no
| 124. yes | no
| 125. yes | no
| 126. yes | no
| 127. yes | no
| 128. yes | no

Specify clinical features:

- Autoimmune hemolytic anemia
- Bone abnormalities
- Edema
- Eosinophilia
- Failure to thrive (weight < 5th percentile)
- Graft versus host disease due to blood transfusion
- Graft versus host disease due to maternal engraftment
- Growth hormone deficiency
- Hepatosplenomegaly
- Hypoproteinemia
- Lymphoproliferative disease
- Maternal T-cell engraftment
- Microcephaly
- Neutropenia
- Skin rash
- Thrombocytopenia (< 100 x 10^9/L)
- Warts
- Other features

Prophylactic Drug Given? Prophylactic Drug Stopped? Date Stopped

<table>
<thead>
<tr>
<th>Prophylactic Drug Given?</th>
<th>Prophylactic Drug Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>129. Antifungal drug(s)</td>
<td>130. yes</td>
<td>131. yes</td>
</tr>
<tr>
<td>2 no</td>
<td>2 no</td>
<td>20</td>
</tr>
</tbody>
</table>

Pre-HSCT Treatment for Immune Deficiency

128. Was treatment given (between diagnosis and prior to the preparative regimen)?

- 1 yes
- 2 no

Complete the table below

Prophylactic drugs paused for < 1 week should not be considered as “Prophylactic Drug Stopped.”

<table>
<thead>
<tr>
<th>Prophylactic Drug Given?</th>
<th>Prophylactic Drug Stopped?</th>
<th>Date Stopped</th>
</tr>
</thead>
<tbody>
<tr>
<td>129. Antifungal drug(s)</td>
<td>130. yes</td>
<td>131. yes</td>
</tr>
<tr>
<td>2 no</td>
<td>2 no</td>
<td>20</td>
</tr>
</tbody>
</table>

Fax this form to your designated campus (Milwaukee 414-805-0714 or Minneapolis 612-627-5895).
CIBMTR Recipient ID: CIBMTR Center Number: 

Therapy Given? Therapy Stopped? Date Stopped
138. Antithymocyte globulin (ATG, ATGAM, Thymoglobulin) 1 yes 139. yes 140. date estimated
2 no 2 no 2

141. Corticosteroids, systemic 1 yes 142. yes 143. date estimated
2 no 2 no 2

144. Corticosteroids, topical 1 yes 145. yes 146. date estimated
2 no 2 no 2

147. Cyclophosphamide (CTX, Cytoxan, Neosar) 1 yes 148. yes 149. date estimated
2 no 2 no 2

150. Cyclosporine (CsA, Neoral, Sandimmune) 1 yes 151. yes 152. date estimated
2 no 2 no 2

153. In vivo monoclonal antibody 1 yes 154. yes 155. yes 156. date estimated
2 no 2 no 2

Specify monoclonal antibody:

154. Alemtuzumab (Campath) 1 yes 155. yes 156. date estimated
2 no 2 no 2

157. Daclizumab (anti-CD25, Zenapax) 1 yes 158. yes 159. date estimated
2 no 2 no 2

160. Etanercept (Enbrel) 1 yes 161. yes 162. date estimated
2 no 2 no 2

163. Infliximab (anti-TNF-α, Remicade) 1 yes 164. yes 165. date estimated
2 no 2 no 2

166. Rituximab (anti-CD20, Rituxan, MabThera) 1 yes 167. yes 168. date estimated
2 no 2 no 2

169. Other monoclonal antibody 1 yes 170. yes 171. date estimated
2 no 2 no 2

172. Specify other monoclonal antibody: 

173. Mycophenolate mofetil (MMF, Cellcept) 1 yes 174. yes 175. date estimated
2 no 2 no 2

176. Tacrolimus (FK506, Prograf) 1 yes 177. yes 178. date estimated
2 no 2 no 2

CIBMTR Form 2031 revision 2 (page 7 of 8) June 2009
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Therapy Given? Therapy Stopped? Date Stopped

179. Other immunosuppressive drug
1 □ yes 180. □ yes 181. □ date estimated
2 □ no

182. Specify other immunosuppressive drug:

183. Was gene therapy performed (between diagnosis and prior to the preparative regimen)?
1 □ yes 184. Specify date of infusion of gene therapy:
2 □ no

[Month Day Year]

185. Was the recipient considered to have failed gene therapy?
1 □ yes
2 □ no

186. Did the recipient receive any other significant treatment(s) (between diagnosis and prior to the preparative regimen)?
1 □ yes 187. Specify other treatment(s): ____________________________
2 □ no

188. Did the patient receive parenteral nutrition (between diagnosis and prior to the preparative regimen)?
1 □ yes
2 □ no

189. Did the patient receive mechanical ventilation (between diagnosis and prior to the preparative regimen)?
1 □ yes
2 □ no

190. Were any biologic specimens collected for this recipient (between diagnosis and prior to the preparative regimen)?
1 □ yes
2 □ no
3 □ unknown

Specify if specimen(s) collected and available for future research:
191. □ yes 2 □ no DNA
192. □ yes 2 □ no Epstein-Barr virus (EBV)-transformed B-cell line
193. □ yes 2 □ no Fibroblast cell line
194. □ yes 2 □ no Herpes virus saimiri-transformed T-cell line
195. □ yes 2 □ no Other T-cell line
196. □ yes 2 □ no Pathological specimen
197. Specify pathological specimen(s):
198. □ yes 2 □ no Peripheral blood mononuclear cells (PBMC), frozen
199. □ yes 2 □ no RNA
200. Specify RNA source: ____________________________
201. □ yes 2 □ no Serum (pre-IVIG)
202. □ yes 2 □ no Other specimen
203. Specify other specimen(s): ____________________________