



**CLINICAL DETAILS (CONT):**

15. Age at presentation: .....

16. Age at definite diagnosis: .....

**16. DIAGNOSTIC CATEGORIES:**

**16.1 Antibody Deficiency:**

Yes  No  Don't know

**If no go to 16.2, if yes please specify:**

- XL agammaglobulinaemia
- Common variable immunodef.
- Hyper IgM syndrome:
- Ig class deficiency

- XL  Sporadic
- Assoc. with thymoma  Assoc. transcobalamin II def
- XL  AR  Sporadic
- Assoc with IgG subclass def (please specify)

- Ig subclass deficiency
- Ig light chain deficiency
- Deficiency of specific antibodies (please specify) .....

Associated with gene deletion

**16.2 T cell and combined deficiencies:  
(T and B cell deficiencies)**

Yes  No  Don't know

**If no go to 16.3, if yes please specify:**

- DiGeorge anomaly
- Ataxia telangiectasia
- Other chromosomal breakage syndromes (please specify) .....
- Wiskott Aldrich syndrome
- T cell receptor deficiency (please specify) .....
- ADA deficiency
- PNP deficiency
- XL severe combined immunodeficiency
- Omenn's syndrome
- Sporadic B cell positive sever combined immunodeficiency
- Other forms of SCID/CID (please specify) .....
- Def HLA expression  def HLA class I + II expression
- def HLA class II expression
- Short limbed dwarfism
- Chronic mucocutaneous candidiasis
- Other (please specify) .....

**16.3 Complement deficiencies**

Yes  No  Don't know

**If no go to 16.4, if yes please specify:**

- C1q  C1r  C1s  C2  C3  C4  C5  C6  C7  C8  C9
- Factor B  Factor D  Properdin
- C1-esterase inhibitor def →  functional  quantitative

16.4 **Phagocytic disorders** →

Yes  No  Don't know

**If no go to 16.5, if yes please specify:**

- Chronic granulomatous disease →  
(please specify subcomponent) .....
- Primary neutropenia (Kostmann's disease)
- LFA deficiency →  LAD I  LAD II
- Chediak-Higashi syndrome
- Congenital asplenia
- Other (please specify) .....

16.5 **Other**

Yes  No  Don't know

**If no go to 17, if yes please specify:**

- X linked lymphoproliferative syndrome
- Hyper IgE syndrome (Job's syndrome)
- Interleukin deficiency (please specify) .....
- Interleukin receptor deficiency (please specify).....
- Any other deficiency (please specify) .....

**INITIAL INVESTIGATIONS:**

- 17. Haemoglobin ..... g/L.
- 18. Platelet count .....x10<sup>9</sup>/L.
- 19. White cell count .....x10<sup>9</sup>/L.
- 20. Absolute lymphocyte count .....x10<sup>9</sup>/L.
- 21. Lymphocyte subset analysis:
  - CD3.....x10<sup>9</sup>/L
  - CD4.....x10<sup>9</sup>/L
  - CD8.....x10<sup>9</sup>/L
  - CD19.....x10<sup>9</sup>/L
- 22. Lymphocyte response to mitogens:
  - specify mitogen .....
  - Absent  Low  Normal  Not done
- 23. Immunoglobulins:
  - IgG.....g/L IgG<sub>1</sub>.....g/L
  - IgG<sub>2</sub>.....g/L
  - IgG<sub>3</sub>.....g/L
  - IgG<sub>4</sub>.....g/L
  - IgA.....g/L
  - IgM.....g/L
- 24. PNP activity .....
- 25. ADA activity .....
- 26. HIV antibody:  Negative  Positive  Not done

**MANAGEMENT:**

27. Duration of care at your centre: .....

28. Is this patient receiving gammaglobulin:  Yes  No  Don't know

29. **If no go to question 34**

**if yes please specify:**

Route of administration:  IV  IM  SC

Frequency of therapy: .....times/year

Amount given on each occasion: .....grams

Duration of therapy at your centre: .....

Usual brand of immunoglobulin: .....

Have they suffered serum sickness as a result of Ig therapy?:  Yes  No  Don't know

Have they contracted hepatitis as a result of Ig therapy?:  Yes  No  Don't know

please specify type if known:.....

Have they suffered any other complication of Ig therapy (please specify) .....

30. Bone marrow transplant performed:  Yes  No  Don't know

31. If yes, Date(s): (1)...../...../....., (2)...../...../....., (3)...../...../.....

Relationship of donor: .....

Donor match ..... out of .....antigens

T-cell depleted marrow  Yes  No

Other tissue (eg; cord blood) .....

Conditioning  Yes  No

32. Lymphokine therapy  
Gamma interferon  Yes  No  
Alpha interferon  Yes  No  
G-CSF  Yes  No  
GM-CSF  Yes  No

33. Other therapy  
C1-esterase inhibitor  Yes  No  
Other (please specify) .....

**OUTCOME INFORMATION**

34.  Alive  with disease  disease free

Died Cause of death if known: .....

**Thank you for completing this questionnaire.**

Please return it in the reply-paid envelope to:

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