Registration process

Different levels of documentation can be chosen for the different disease groups (e.g. a centre chooses level 2 only for Antibody deficiencies and Complement deficiencies and level 1 for all others). Level 3 forms will be added later for dedicated studies.

There are two basic types of forms:

• **Global forms:**
  – exist only once per patient
  – can also be list forms containing several entries

• **Date-dependent forms (=current data forms)**
  – exist once per documentation date
Relevant Forms Level 1

• Global forms:
  – Patient info
  – Way to Diagnosis
  – PID Diagnosis (list)
  – HSCT and Gene Therapy (lists)
  – Death report form

• Date-dependent forms (=current data forms):
  – Current status
  – Current treatment:
    • Ig-Replacement
Relevant Forms Level 2

• Global forms:
  – Patient info
  – Way to Diagnosis
    + Clinical Manifestations
    + Laboratory values
  – PID Diagnosis (list)
  – HSCT and Gene Therapy (lists)
    + possibly further details on HSCT and gene therapy
  – Death report form
  – Malignancies (list)

• Date-dependent forms (=current data forms)
  – Current status
  – Current clinical manifestations
  – Quality of Life
  – Current laboratory values
  – Current treatment:
    • Ig-Replacement
    • Other treatments (antibiotics, immunosuppressives, PEG-ADA, G-CSF…)

18.10.2013
Data forms and levels

Level 1

- Patient Info
- Way to diagnosis + PID Diagnosis
- Current status
- Any changes to Diagnosis, any new HSCT or gene therapy?

Level 2

- Clin. manifestations from birth to dx
- Lab values leading to initial dx
- Current Therapy
- Current lab values
- QoL
- Clin. manifestations
- Malignancies

To be documented at baseline only

To be documented at baseline and every follow-up

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• **Level-1 Documentation**
  – At initial registration: Basic diagnostic data, HSCT/gene therapy and current Ig replacement at initial registration
    **AND**
  – In every subsequent calendar year: Changes to living status or diagnosis, HSCT/gene therapy/ Ig replacement update

• **Level-2 Documentation**
  – At initial registration: Complete diagnostic data, current therapy, QoL; if diagnosis > 12 months ago: current Clinical manifestations, lab values
    **AND**
  – In every subsequent calendar year: Current clinical manifestations, lab values, therapy, QoL (only if news are available)

• **Comments:**
  – For deceased patients, the “death report form” must be filled in

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The following flowcharts describe the documentation process for each documenting centre
Patient visit

Start → Patient visit → Patient registered?
  yes → Level? 2
  no → Get informed consent

Have you already reported on this patient in the current year?
  yes → Fill in Level 1 Current data forms
  no → Level? 1

Fill in Level 1 global forms

Level? 2

Fill in Level 2 "global" forms, QoL, current therapy form (incl. Ig repl.)

Fill in Level 2 Clin. Manifestations & lab values

Diagnosis made > 12 months ago
  yes → Fill in Level 2 Clin. Manifestations & lab values
  no → Fill in current Ig replacement

Stop

18.10.2013
Dear Dr. ABC,
Patients x, y, z have not been documented yet this year

For each patient

Level?

1 Fill in Level 1 Current data forms

2 Fill in Level 1+2 Current data forms

Stop
Option “patient questionnaire”
For new calendar year (level 2)

Start

January 1st

Download questionnaire for each patient from the registry

Put questionnaires into patient files

Stop