This form shall be filled in when the patient is **updated (once a year).**
If data from the patient **has been transferred from the old registry**, the baseline form must be completed first. The follow-up short form shall then be used starting the year after.

Follow-up short form

Date of documentation: [Date]	<<This date is automatically displayed and stored by the system (based on the server clock)>>

Date of last visit or last news

Date of last news from patient: [Date] ☐ No news from patient since last documentation	<< Can only store a date that comes after the most recent documentation date. >> Enter the date when you last received news on the patient. If you have **no news at all** from the patient, please select 'No news from patient since last documentation'.
Type of news: ☐ last clinical visit ☐ last news (phone, letter etc.) ☐ unknown	Indicate whether this is the date of the last patient visit to the physician or hospital, or whether information was received e.g. by telephone from the patient, the patient's family or a physician.

All following questions refer to the **timepoint of the patient's last visit or date of last news**.

Current status: ☐ Alive ☐ Deceased ☐ Lost to follow-up ☐ Discharged after complete recovery	Indicate whether the patient is still alive based on the latest information you have. If you have no information on the current status of the patient because you have lost contact to him/her and do not expect to hear from him/her again, select "lost to follow-up". If the patient is not followed anymore because the immunodeficiency has resolved, select 'discharged after complete recovery'. << If "deceased" is selected, the "Death report form" will open. Centres will not be asked to update patients with status "deceased", "lost to follow-up" and "Discharged" afterwards >>
Details on status: _____	<<optional. Appears if "Lost to follow-up" is selected>> If the patient is lost to follow-up, you may enter details here (e.g. name of physician the patient was referred to)

Changes to diagnosis

Changes to diagnosis: ☐ PID diagnosis has changed ☐ No PID after all ☐ No change << default value >>	If the PID diagnosis has changed or if you have identified the affected gene, select "PID diagnosis has changed" and fill in the respective fields. If the PID diagnosis has been incorrect and there is no PID in this patient at all, please select "No PID after all".
Current PID Diagnosis << new entry >>	<<only available if "PID diagnosis has changed" has been selected. If the affected gene has been identified, this is stored with the existing entry. Otherwise, the system creates a completely new PID Diagnosis entry (thereby creating a "diagnostic history")>>

	Select the most recent PID diagnosis for this patient (corresponding to the most recent visit date or date of last news). If you cannot find the appropriate disease, or if a disease is missing, send an email to esid.registry@kenes.com.
Affected gene: [List of genes] <<or>> ☐ Genetically tested, but no mutation found ☐ Not genetically tested ☐ History of genetic tests unknown	Select the gene, in which disease-causing mutation(s) have been found in this patient. If you have sequenced one or more of the known genes but have found no mutation, select "no mutation found". If no molecular analysis has been performed at all, select "not genetically tested". If a gene is missing, send an email to esid.registry@kenes.com.
Additional genes: _____	If more than one PID-causing gene mutation has been found in this patient, or if other gene mutations have been found, you can enter these here.
Date of genetic diagnosis: [Year] [Month] [Day] ☐ unknown	<<Will only be visible if an affected gene has been selected. Cannot be previous to date of birth. Exception: If previous to date of birth less than 9 months, form can only be stored if "Prenatal diagnosis" in the next field is selected. >> If applicable, enter the date when the genetic diagnosis was confirmed (date of molecular analysis). If month and/or day are unknown, leave them open.
Sequencing method: ☐ Gene sequencing ☐ Whole exome/genome sequencing ☐ Non-genetic definitive test ☐ Unknown	<<Will only be visible if an affected gene has been selected>> Select the sequencing method applied. If the molecular analysis was performed as a candidate gene testing (using the "traditional" method of Sanger sequencing), select "Gene sequencing". If the whole genome or exome was sequenced, select that option. If a non-genetic test like 22q11 FISH for DiGeorge syndrome was used, select 'Non-genetic definitive test'.
Lab that performed the genetic analysis: <<Drop down list>>	<<Only visible if a gene or "no mutation found" has been selected >> If the lab that performed the analysis is not in the list, send an email to esid.registry@kenes
Genetics laboratory: <<Drop down list>>	<<Only visible if a gene has been selected >> Select the name of the centre that performed the genetic analysis.
Reason for genetic analysis: ☐ Analysis following clinical diagnosis ☐ Family screening ☐ Prenatal diagnosis ☐ Diagnosis by neonatal screening ☐ unknown	<<Will only be visible if an affected gene has been selected. Only one can be selected>> Select the reason for the molecular analysis in this patient.

Stem cell / gene therapy

Has one of the following been performed in this patient since the last documentation?	
Haematopoietic stem cell transplantation (HSCT): ☐ Yes ☐ No ☐ Unknown	<< Defaults to "yes" if "yes" has been selected in the previous documentation timepoint.>> Indicate whether haematopoietic stem cell transplantation (HSCT) has been performed in this patient since the last documentation.
If yes, SCETIDE ID: **EBMT ID:**	If the patient has been reported to SCETIDE, provide the ID number. SCETIDE is the registry for Stem Cell Transplants for primary Immune Deficiencies in Europe, in collaboration with ESID. Contact info.scetide@nck.aphp.fr for more information. If the patient has been reported to the EBMT Registry, provide the ID number.
If yes, enter for each transplantation:	<<several entries possible. Previous entries are listed and do not have to be entered again. >>
Date of transplantation [Year] [Month] [Day] ☐ unknown	<<Cannot be previous to the date of birth. >> Please enter the date of the HSCT.
Type of donor ☐ MSD (Matched sibling donor) ☐ MUD (Matched unrelated donor) ☐ MMUD (Mismatched unrelated donor) ☐ Haplo-identical (parent) donor ☐ Autologous ☐ Other related donor ☐ Unknown	Indicate the type of stem cell donor.
Source of CD34 stem cells ☐ bone marrow ☐ peripheral blood ☐ cord blood ☐ unknown	Indicate the source of the stem cells used in this HSCT.
Gene therapy ☐ Yes ☐ No ☐ unknown	<< Defaults to "yes" if "yes" has been selected in the previous documentation timepoint.>> Indicate whether gene therapy has been performed in this patient since the last documentation.
If yes, enter for each gene therapy:	<<several entries possible. Previous entries are listed and do not have to be entered again. >>
Date of gene therapy [Year] [Month] [Day] ☐ unknown	<<Only visible if "yes" has been selected in previous question. Cannot be previous to the most recent visit date. >> Enter the date when the gene therapy was initiated.

Immunoglobulin (Ig) replacement

Does the patient currently receive Ig-replacement? ☐ Yes ☐ No ☐ unknown	
Current brand name [List of brand names, unknown]	Which brand of immunoglobulins does the patient currently use?
Current route of administration ☐ intravenous ☐ subcutaneous ☐ intramuscular	<<Route is automatically selected by the system if a respective brand name is selected>>
Current place of administration ☐ home ☐ hospital ☐ hospital: inpatient ☐ hospital: outpatient ☐ both (home&hospital) ☐ unknown	Select the place of administration of Ig replacement. If the infusion is given in a hospital, indicate whether this is done during an inpatient or outpatient stay. If this is not known, choose the option "hospital".
Current dose [integer] mg/kg body weight or [decimal] ☐ g ☐ ml ☐ dose unknown **Interval for this dose** Every [integer] ☐ week(s) ☐ day(s) <<or>> [integer] times per ☐ week ☐ month ☐ year ☐ interval unknown	Indicate the current dose and frequency of Ig replacement in this patient. Enter the relative dose (per kg body weight) or the absolute dose, or both, if available. Enter the **actual interval**, e.g. if the patient applies 10 ml of SCIg every second day, enter "10 ml every 2 days", and NOT "35 ml every week". In the case of **alternating doses** (e.g. "10 ml one week and 15 ml the next week"), calculate the mean value (e.g. "12.5 ml every week").
Patient's current weight [integer] kg ☐ unknown	<< If brand name, weight, and absolute dose are entered, the system will calculate and store the mg/kg dose. If the mg/kg is entered manually in addition and differs from the calculated value, the system asks the user which value should be used.>>
Current side effects ☐ Yes ☐ No ☐ unknown	Indicate whether the administration of the current Ig replacement has ever caused or is still causing side effects.
If yes, type of side effects <<one or several possible>> ☐ anaphylaxis ☐ fever ☐ headache ☐ local side effects ☐ renal failure ☐ aseptic meningitis ☐ venous thrombosis ☐ arterial thrombosis ☐ Other, specify: _____	<<Only visible if "side effects" has been answered with "yes">>