**Supplementary Table-1 (Card A). Initial patient information**

|  |  |
| --- | --- |
| Full name |  |
| Year of fulfillment |  |
| Date of birth |  |
| Diagnosis of immunodeficiency (descriptively + ICD-10) |  |
| Date of the diagnosis of deficiency + age at which the deficiency was diagnosed | Date: Age: |
| Year of the occurrence of the first symptoms + age when deficiency symptoms first appeared | Year: Age: |
| Comorbidities |  |
| Allergies (including information about intolerance to any immunoglobulin preparation) |  |
| Immunoglobulins that are administered to the patient (trade name, dose per kg of body weight, dose per month)  Note: If the patient does not receive immunoglobulins, write: not applicable |  |
| Other drugs |  |
| Name and stamp of the center where the patient is treated |  |
| Name of the doctor who is treating the patient |  |
| Phone number of the center where the patient receives care  ● Secretary’s office  ● Nurses’ station  ● Doctor's office |  |
| Stimulants |  |
| Height (cm) |  |
| Bodyweight (kg) |  |
| BMI and nutritional status score: malnutrition(<18,5)/normal nutrition(18,5-24,9)/overweight(25,0-29,9)/obesity(≥30) |  |
| Vaccinations (in the case of indications or contraindications to vaccinations, this should also be included in this point, as well as information on the vaccinations received thus far) |  |
| Other important information |  |

**Supplementary Table-2 (Card B). Card of initial examinations and consultations—qualifying (performed during the diagnostic process and/or in the first year after the diagnosis)**

|  |  |  |
| --- | --- | --- |
| **Full name:** | | |
| **Current year:** | | |
| **Examination** | **Examination result (N/A)** | **Comments** |
| **Initial physical examination** | | |
| Bodyweight |  |  |
| BMI | Result: \_\_\_  malnutrition(<18,5)/normal nutrition(18,5-24,9)/overweight (25,0-29,9)/obesity(≥30) | Underline as appropriate |
| BP+HR |  |  |
| Oral cavity |  |  |
| Nasal cavity |  |  |
| Ears |  |  |
| Lungs |  |  |
| Abdomen | Liver  Spleen  Others |  |
| Lymph nodes |  |  |
| Skin |  |  |
| Others (please specify) |  |  |
| **Initial laboratory tests** | | |
| Complete blood count with manual differential |  |  |
| Creatinine1 |  |  |
| GFR1 |  |  |
| Glucose1 |  |  |
| AST1 |  |  |
| ALT1 |  |  |
| Alkaline phosphatase1 |  |  |
| GGT1 |  |  |
| LDH1 |  |  |
| β2-Microglobulin1 |  |  |
| ESR1 |  |  |
| CRP1 |  |  |
| Proteinography1 |  |  |
| Calcium1 |  |  |
| VitaminD31 |  |  |
| Vitamin B121 |  |  |
| Folic acid1 |  |  |
| Transferrin/ferritin1 |  |  |
| Uric acid1 |  |  |
| TSH andFT41 |  |  |
| Lipid profile1 |  |  |
| General urine test1 |  |  |
| Fecal occult blood test1 |  |  |
| HBS1 |  |  |
| Tumor markers1or3 |  |  |
| INR1 |  |  |
| APTT1 |  |  |
| IgA1 |  |  |
| IgG1 |  |  |
| IgM1 |  |  |
| IgG subclass1 |  |  |
| IgE1 |  |  |
| EBV PCR1or3 |  |  |
| CMV PCR1or3 |  |  |
| HCV PCR1or3 |  |  |
| HIV PCR1or3 |  |  |
| Others (e.g.serum amyloid A – SAA) |  |  |
| **Initial radiological examination** | | |
| Chest radiography/lung ultrasound1 |  |  |
| Chest CT/MRI2 |  |  |
| Abdominal ultrasound1 |  |  |
| Abdominal CT/MRI2 |  |  |
| Sinuses CT/MRI2 |  |  |
| Ultrasound of the lymph nodes1 |  |  |
| Thyroid ultrasound2 |  |  |
| Breast ultrasound/MRI\*,2 |  |  |
| Gynecologic ultrasound\*,2 |  |  |
| Testicular ultrasound\*\* |  |  |
| Echocardiography2 |  |  |
| Others: |  |  |
| **Initial endoscopic examination** | | |
| Gastroscopy2 |  |  |
| Colonoscopy2 |  |  |
| Bronchoscopy3 |  |  |
| Others |  |  |
| **Other examinations** | | |
| Spirometry1 |  |  |
| DLCO2 |  |  |
| Lymphocyte phenotyping1 |  |  |
| Fecal occult blood test2 |  |  |
| Microbiologic examination3 (e.g. parasite screening) |  |  |
| Cytology\* |  |  |
| Others: e.g.body plethysmography |  |  |
| **Consultations** | | |
| Dental2 |  |  |
| Ophtalmology2 |  |  |
| Otolaryngology2 |  |  |
| Gynecologic\*,2 |  |  |
| Urologic\*\*\*,2 |  |  |
| Psychological1 |  |  |
| Psychiatric1 |  |  |
| Dermatology1 |  | Indicated evaluation of moles |
| Others: |  |  |

1. Should be performed at the time of diagnosis
2. Should be performed within the first year after diagnosis
3. Decision about the examination/consultation belongs to the treating physician, in the absence of indications, please write: currently no clinical indications

\*applies to women

\*\*applied to men

\*\*\*applies to all men and women with urinary symptoms

Proposed abbreviations (physicians can fill in the table with these keywords, they were suggested for ease and standardization):

(N – the normal result, A - the abnormal result, NA - not applicable, CNCI - currently no clinical indications, DNC - did not consent, ITP - impossible to perform, e.g. due to contraindications or the patient's inability to perform, NPB - not performed before)

**Supplementary Table-3 (Card C) Card of control examinations and consultations**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Full name:** | | | | | | | | |
| **Current year:** | | | | | | | | |
| **Examination** | **Date and result I**  **(N/A)** | | | **Date and result II**  **(N/A)** | | **Recommended frequency** | | **Comments** |
| **Physical examination (check-up)** | | | | | | | | |
| Bodyweight |  | | |  | | Group 1 (Table A)  -at least once a year Group 2  At least twice a year | |  |
| BMI |  | | |  | | as above | | Specify:malnutrition(<18,5)/ normal nutrition(18,5-24,9)/ overweight(25,0-29,9)/ obesity(≥30) |
| BP+HR |  | | |  | | as above | |  |
| Oral cavity |  | | |  | | as above | |  |
| Nasal cavity |  | | |  | | as above | |  |
| Ears |  | | |  | | as above | |  |
| Lungs |  | | |  | | as above | |  |
| Abdomen | Liver  Spleen  Others | | |  | | as above | |  |
| Lymph nodes |  | | |  | | as above | |  |
| Skin |  | | |  | | as above | |  |
| Others (please specify) |  | | |  | |  | |  |
| **Follow-up laboratory tests** | | | | | | | | |
| Complete blood count with differential |  | | |  | | Group 1 (Table A)  -at least every 1-2 years  Group 2  -at least twice a year | |  |
| Creatinine |  | | |  | | as above | |  |
| GFR |  | | |  | | as above | |  |
| AST |  | | |  | | as above | |  |
| ALT |  | | |  | | as above | |  |
| Alkaline phosphatase |  | | |  | | as above | |  |
| LDH |  | | |  | | as above | |  |
| Glucose |  | | |  | | as above | |  |
| ESR |  | | |  | | as above | |  |
| CRP |  | | |  | | as above | |  |
| General urine test |  | | |  | | as above | |  |
| Uric acid |  | | |  | | Group 1 (Table A)  -at least every 1-2 years  Group 2  -at least once a year | |  |
| Lipid profile |  | | |  | | as above | |  |
| Sodium |  | | |  | | as above | |  |
| Potassium |  | | |  | | as above | |  |
| Proteinography |  | | |  | | as above | |  |
| β2-microglobulin |  | | |  | | as above | |  |
| Fecal occult blood test |  | | |  | | as above | |  |
| TSH |  | | |  | | At least every 1-3 years | |  |
| Calcium |  | | |  | | as above | |  |
| Vitamin D3 |  | | |  | | as above | | It is best to determine its concentration after the summer season |
| APTT |  | | |  | | as above | |  |
| INR |  | | |  | | as above | |  |
| Tumor markers1 |  | | |  | | as above | |  |
| Folic acid |  | | |  | | as above | | Additionally, in the case of anemia, malabsorption and polyneuropathy |
| Vitamin B12 |  | | |  | | as above | |
| Transferrin/ferritin |  | | |  | | as above | |
| HBS |  | | |  | | Indicated in clinically justified situations | |  |
| EBV PCR1 |  | | |  | |  |
| CMV PCR1 |  | | |  | |  |
| HCV PCR1 |  | | |  | |  |
| HIV PCR1 |  | | |  | |  |
| IgG |  | | |  | | Group1  - at least every 1-3 years  Group 2  -IVIG- it is advisable to test before the next transfusion  - SCIG - once every 3-6 months | | Determining the main classes of immunoglobulins, despite disturbing their production, can help in the early detection of the presence of a pathological protein, e.g. a monoclonal protein |
| IgG subclass |  | | |  | | Group 1  -at least every 1-3 years  Group 2  -indicated in clinically justified situations | |
| IgA |  | | |  | | At least every 1-3 years | |
| IgM |  | | |  | | At least every 1-3 years | |
| Others ( e.g.serum amyloid A – SAA) |  | | |  | |  | |  |
| **Follow-up radiological examination** | | | | | | | | |
| Examination  (circle the appropriate examination) | | | Date and result of the examination (N/A) | | Date of the next examination | | Comments | |
| Chest radiography/lung ultrasound2 | | |  | |  | | At least every 2 years (Group 1)  1-2 times a year (Group 2) | |
| Chest CT/MRI2 | | |  | |  | | CT (preferably HRCT) is recommended every 2-4 years (at least once every 5 years) in patients with documented and potentially progressive abnormalities in the lungs. At least 1 time in 5 years in patients with normal initial result (Group 2). In the case of a large number of CT examinations, MR is indicated | |
| Abdominal ultrasound2 | | |  | |  | | At least every 2 years (Group 1)  At least once a year (Group 2) | |
| Abdominal CT/MRI2 | | |  | |  | | At least 1 time in 5 years in patients with normal initial result (Group 2). In the case of a large number of CT examinations, MR is indicated | |
| Sinuses CT/MRI2 | | |  | |  | | 1 time in 5 years or less if the initial result is correct ( Group2) | |
| Ultrasound of the lymph nodes2 | | |  | |  | | At least every 2-4 years | |
| Thyroid ultrasound2 | | |  | |  | | Optimally once a year, at least once every 2 years | |
| Breast ultrasound/MRI\*,2 | | |  | |  | | Optimally once a year | |
| Gynecologic ultrasound\*,2 | | |  | |  | | Optimally once a year, at least once every 2 years | |
| Testicular ultrasound\*\*,2 | | |  | |  | | Screening every 3 years | |
| Echocardiography2 | | |  | |  | | At least every 5 years, and every 2-4 years in the case of mild cardiovascular diseases | |
| Others: | | |  | |  | |  | |
| **Follow-up endoscopic examination** | | | | | | | | |
| Examination | | Date and result of examination (N/A) | | | Date of the next examination | | Comments | |
| Gastroscopy2 | |  | | |  | | At least every 3 years | |
| Colonoscopy2 | |  | | |  | | At least every 3 years | |
| Bronchoscopy4 | |  | | |  | |  | |
| Others: | |  | | |  | |  | |
| **Other follow-up examination** | | | | | | | | |
| Examination | | Date and result of examination (N/A) | | | Recommended frequency | | Comments | |
| Spirometry | |  | | | At least once a year | |  | |
| DLCO | |  | | | At least once a year | |  | |
| Lymphocyte phenotyping | |  | | | At least every 1-3 years | |  | |
| Microbiologic tests  (e.g.parasite screening) | |  | | | As needed, depending on the clinical condition | |  | |
| Cytology\* | |  | | |  | |  | |
| Others: e.g.body plethysmography, densitometry | |  | | |  | |  | |
| **Consultations** | | | | | | | | |
| Type of consultation | | Date and result of examination (N/A) | | | Recommended frequency | | Comments | |
| Dental | |  | | | At least twice a year | |  | |
| Ophthalmology | |  | | | At least every 1-3 years | |  | |
| Otolaryngology | |  | | | At least every 1-3 years | |  | |
| Rehabilitation | |  | | | At least once a year | | To teach and control pulmonary rehabilitation | |
| Gynecologic\* | |  | | | At least once a year | |  | |
| Urologic\*\*\* | |  | | | At least once a year | |  | |
| Dermatology | |  | | | At least once a year | | Evaluation of moles | |
| Psychological1 | |  | | |  | |  | |
| Psychiatric1 | |  | | |  | |  | |
| Oncology1 | |  | | |  | |  | |
| Others: | |  | | |  | |  | |

1. Decision about the examination/consultation belongs to the treating physician, in the absence of indications, please write: currently no clinical indications
2. Routine frequency of examinations described in the text, additionally indicated in clinically justified situations

\*applies to women

\*\*applied to men

\*\*\*applies to all men and women with urinary symptoms

Proposed abbreviations (physicians can fill in the table with these keywords, they were suggested for ease and standardization):

(N – the normal result, A - the abnormal result, NA - not applicable, CNCI - currently no clinical indications, DNC - did not consent, ITP - impossible to perform, e.g. due to contraindications or the patient's inability to perform, NPB - not performed before)