



CORE Dataset: USIDNET Immunodeficiency Patient Registry

Patient name: _____ Registry ID Number: _____

This CORE questionnaire is to be completed for all patients, regardless of diagnosis

Patient Consent: Yes: Institution specific IRB [] Western IRB []

IRB waiver [] Specify reason _____

Has the patient agreed to allow USIDNET Registry personnel to contact him/her? Yes / No

Enter date of most recent visit (mm/dd/yyyy) ____/____/____

Enter date this Record was prepared (mm/dd/yyyy): ____/____/____

Enter dates covered in this form (mm/dd/yyyy): ____/____/____ to ____/____/____

Primary Immunodeficiency Diagnosis: Circle appropriate diagnosis

Severe Combined Immunodeficiency (SCID): X-linked (γ c), ADA, JAK3, RAG1-2, Artemis, IL-7R α (CD127), CD45, CD3 δ , CD3 ϵ , CD3 ζ , Omenn, other _____

Combined Immunodeficiency (CID): Bare lymphocyte (MHC class-II deficiency), PNP, ZAP70, CD25 (IL2- R α), Cartilage-Hair Hypoplasia, MHC class-I deficiency, other _____

Humoral deficiencies: Common Variable Immunodeficiency (CVID), X Linked Agammaglobulinemia (XLA), AR agammaglobulinemia, Selective IgA deficiency, IgG subclass deficiency, Specific antibody deficiency, other _____

Hyper IgM syndrome (HIGM): X-linked HIGM (CD40L), CD40L, CD40, UNG, AID, other _____

Chronic Granulomatous Disease (CGD): X-linked (gp91phox), p47phox, p22phox, p67phox, unspecified _____

Leukocyte Adhesion Deficiency (LAD): type 1 (CD18), type 2 (FUCT1), type 3 (CalDAG-GEFI), other _____

Complement deficiency: C1q (α, β, γ), C1r, C1s, C2, C3, C4A, C4B, C5, C6, C7, C8 $\alpha-\gamma$, C8 β , C9 deficiency, Factor B, Factor D, Factor H, Factor I, Properdin, C1 esterase inhibitor, other _____

Syndromes: Circle appropriate syndromes

Wiskott-Aldrich Syndrome (WAS):
(classic, XLT, XL Neutropenia)

ICF (immunodeficiency, centromere instability and facial anomalies) syndrome

DiGeorge Syndrome (DGS)

IPEX (Immune dysregulation, polyendocrinopathy, enteropathy, X-linked syndrome)

Chediak-Higashi syndrome

NBS (Nijmegen Breakage Syndrome)

Chronic Mucocutaneous Candidiasis

X Linked Lymphoproliferative Syndrome (XLP)

Ataxia Telangectasia

NEMO

Hyper IgE syndrome (Dominant, STAT3)(recessive)

Other (explain): _____

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Patient Name, Initials or Site Study Participant #: _____

Is this the initial registration of this patient [] or follow-up? []

Patient Identification:

Requested data shown in **BOLD** on this page should **not be provided** without signed patient consent or IRB waiver.

Site assigned Identification/Study Participant # (if applicable): _____

Patient Name:

First: _____ **Middle:** _____ **Last:** _____

Maiden/Other Name (that may be listed in Registry): _____

Patient Initials ___/___/___ **Date of Birth** ___/___/___ (mm/dd/yyyy) or Birth Year _____

Gender: male [] female []

Home address: (Bold address areas optional for all patients. Include State of Residence for all patients)

Street Address: _____

City: _____

State: _____

Zip Code: _____

Phone: _____

Email: _____

State or Province of birth: _____

Country of birth: _____

Physician or Center Enrolling this Patient:

Specialty of Physician: _____

Name: _____

First: _____

Last: _____

Address: _____

City, State: _____

Zip Code: _____

Phone: _____

Email: _____

Fax: _____

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Patient Name, Initials or Site Study Participant #: _____

Name and role of person completing this data entry form: -

(Resident physician, nurse practitioner, student, study coordinator, principal investigator, etc.)

Other Physicians following this patient: please list additional physicians in the Memo section.

Name:	First:	Last:
Specialty		
Address:		
City, State:		
Zip Code:		
Phone:		
Email:		
Fax:		

Specialty of Physician making the diagnosis of primary immunodeficiency in this patient:-

Specialty of Physician: _____

Name:	First:	Last:
Address		
Phone		
Email		

Race/Ethnicity:

Is the patient Hispanic or Latino (a person of Cuban, Mexican, South or Central American, or other Spanish culture or origin, regardless of race)? Yes [] No []

Is the patient (check all that apply)?

Caucasian/white [] African American/ Black []
Asian or Pacific Islander [] Some Other Race: [] Specify: _____
American Indian/Native American []

Family History:

Familial case: Are there other patients with primary immunodeficiency in the family? Yes [] No [] Unknown []

If yes: How many relatives also have Primary Immunodeficiency? []

Relationship	Diagnosis:	Listed in the Registry?
_____	_____	Y [<input type="checkbox"/>], N [<input type="checkbox"/>]
_____	_____	Y [<input type="checkbox"/>], N [<input type="checkbox"/>]
_____	_____	Y [<input type="checkbox"/>], N [<input type="checkbox"/>]

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Consanguinity (Are the parents of the patient related to each other):

[] Yes (explain) _____ [] No [] Unknown

Did a positive family history prompt testing of this patient? Y [] N []

Patient's height

(age: ___ yrs ___ mos or date ___/___/___)(mm/dd/yyyy) _____ft _____ in or _____cm

Patient's weight

(age: ___yrs ___mos or date ___/___/___)(mm/dd/yyyy) _____lb _____oz or _____ kg

Head circumference:

(age: ___yrs ___mos or date ___/___/___)(mm/dd/yyyy) _____ in or _____cm

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Onset of Symptoms: First severe infection or other characteristic manifestation of the respective PIDD:

Age: ____ years &/or _____ months or date ___/___/_____(mm/dd/yyyy).

Diagnosis established: age ____ years &/ or _____ months or date ___/___/_____(mm/dd/yyyy)

What were the patient's Symptoms/Clinical Problems?

For each issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A Prominent feature for this patient

At initial visit please report findings at or before diagnosis (A) in addition to findings occurring more recently (B), or/& (C) below.

Infections: [] section skipped

If causative organism is known, please indicate corresponding number from table to the right. If other, please specify. Please indicate all that apply.

	A	B	C	Organism(s) (if known)
Abscess:				
Brain				
Liver				
Lung				
Peri-rectal				
Renal				
Skin				
Spleen				
Other:				
Cellulitis				
Conjunctivitis				
Dermatitis (fungal)				
Diarrhea (infectious)				
Empyema				
Encephalitis				
Fever				
Genital				
Hepatitis				
Impetigo				
Lymphadenitis				
Mastoiditis				
Meningitis				
Omphalitis				
Osteomyelitis				
Otitis media				
Peritonitis				
Pharyngitis				
Pneumonia				
Proctitis				
Pyoderma				
Septic arthritis				
Sepsis				
Sinusitis				
Skin				

	List of Organisms
1	Adenovirus
2	Aspergillus
3	Candida
4	Chlamydia
5	Clostridium difficile
6	CMV
7	Coxsackie
8	Cryptococcus
9	Cryptosporidium
1 0	E. coli
1 1	Enterovirus
1 2	Epstein-Barr virus (EBV)
1 3	Giardia
1 4	Haemophilus influenzae
1 5	Herpes simplex
1 6	HHV8 (human herpes virus 8)
1 7	Histoplasmosis
1 8	HIV
1 9	Influenza
2 0	Measles
2 1	Molluscum
2 2	Mycobacterium tuberculosis
2 3	Mycobacteria (nontuberculous)
2 4	Mycobacteria (atypical)
2 5	Mycoplasma
2 6	Neisseria gonorrhoea
2 7	Neisseria meningitides Type_____
2 8	Nocardia
2 9	Papilloma virus (warts)
3 0	Parainfluenza
3 1	Parvovirus B19
3 2	Pneumocystis jiroveci (carinii)
3 3	Polio (wild type)

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	A	B	C	Organism(s) (if known)
Thrush				
Tonsillitis				
Urinary tract infection				
Cystitis				
Pyelonephritis				
Prostatitis				
Urethritis				
Other infection: Specify:				

		List of Organisms
3	4	Polyoma virus (JC, BK)
3	5	Pseudomonas
3	6	Rotavirus
3	7	RSV
3	8	Rubella
3	9	Salmonella
4	0	Serratia
4	1	Shigella
4	1	Staphylococcus aureus
4	2	Staphylococcus aureus (MRSA)
4	3	Streptococcus pneumoniae
4	4	Varicella- primary
4	5	Varicella reactivation
		Other? Specify:
		Other? Specify:

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Systemic or organ specific conditions

For each indicated issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A frequent feature for this patient

Autoimmunity: no change from previous entry section skipped None of the following

	A	B	C
Dermatomyositis			
Henoch-Schonlein Purpura (HSP)			
Iritis			
Systemic lupus erythematosus (SLE)			
Scleroderma			
Uveitis			

	A	B	C
Vasculitis- CNS			
Vasculitis-large vessel			
Vasculitis- small vessel			
Other?			

Cardiovascular no change from previous entry section skipped None of the following

	A	B	C
Congenital heart disease			
Constrictive pericarditis			
Hypertension			
Myocardial infarction			
Stroke (CVA)			
Thrombosis			
Transient ischemic attack			
Vascular malformation			
Other? Specify:			

Constitutional no change from previous entry section skipped None of the following

	A	B	C
Aches			
Fatigue			
Fever			
Malaise			
Sleeplessness			
Sleepiness			
Sweating			
Other? Specify:			

Dental/Oral no change from previous entry section skipped None of the following

	A	B	C
Aphthous ulcers			
Caries (excessive)			
Cleft palate			
Delayed eruption of teeth			
Delayed shedding of teeth			
Gingivitis			
Hypodontia			
Abnormal tooth morphology			
Missing teeth			
Cone/Peg teeth			
Other?			

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For each indicated issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A frequent feature for this patient

Endocrine – Metabolic: no change from previous entry section skipped None of the following

	A	B	C
Adrenal insufficiency			
Cushing's disease			
Diabetes-type 1			
Diabetes-type 2			
Growth hormone deficiency			
Hashimoto's thyroiditis			
Hypercalcemia			
Hypocalcemia			

	A	B	C
Hyperthyroidism			
Hypothyroidism			
Hyperparathyroidism			
Hypoparathyroidism			
Primary ovarian failure			
Pituitary insufficiency			
Other?			

Gastrointestinal:: no change from previous entry section skipped None of the following

	A	B	C
Abdominal pain			
Aphthous ulcers			
Appendicitis			
Autoimmune hepatitis			
Celiac disease			
Cirrhosis			
Inflammatory Bowel Disease			
Colitis			
Enteritis			
Constipation			
Diarrhea (Intermittent)			
Diarrhea (Chronic)			
Eosinophilic esophagitis			
Fistula			
Gall stones			
Gall bladder disease			

	A	B	C
GE reflux			
Hepatomegaly			
Intestinal nodular lymphoid hyperplasia			
Liver failure			
Liver function abnormalities			
Malabsorption			
Obstruction			
Perianal ulceration			
Peritonitis			
Proctitis			
Protein losing gastroenteropathy			
Sclerosing cholangitis			
Other? Specify:			

Genitourinary: : no change from previous entry section skipped None of the following

	A	B	C
Cystitis (Non-infectious)			
Epididymitis			
Glomerulonephritis			
Hemolytic uremic syndrome			
Hematuria			
Nephritis (unspecified)			
Nephrosis			
Orchitis			
Prostatitis			
Renal stones			
Renal failure			
Urinary outlet obstruction			
Other?			

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For each indicated issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A frequent feature for this patient

Growth and development: no change from previous entry section skipped None of the following

	A	B	C
Delayed developmental milestones			
Dysmorphic features			
Failure to thrive or unintended weight loss			
Mental retardation			
Over weight			
Short stature			
Under weight			
Other?			

Hematologic-Lymphoid: no change from previous entry section skipped None of the following

	A	B	C
Absent lymph nodes			
Absent thymus shadow			
Absent tonsils			
Anemia			
Aplastic			
Chronic infection			
Hemolytic anemia			
Immune			
Iron deficiency			
Pernicious			
Unspecified			
Coagulopathy			
Blood transfusion reaction			
Engraftment by maternal cells			
Epistaxis			
Granulomas			

	A	B	C
Hemarthrosis			
Hypersplenism			
Leukocytosis			
Lymphadenopathy			
Lymphopenia			
Myelodysplasia			
Neutropenia			
Petechia			
Splenomegaly			
Thrombocytopenia			
Thrombocytopenia-immune			
Thrombosis			
Thrombotic thrombocytopenic purpura (TTP)			
Other?			

Musculoskeletal : no change from previous entry section skipped None of the following

	A	B	C
Arthritis			
Osteo			
Rheumatoid			
Juvenile Rheumatoid arthritis			
Unspecified			
Arthralgia			
Frequent fractures			
Hemarthrosis			
Hyperextensibility			
Myositis			
Osteoporosis			
Scoliosis			
Short stature			
Skeletal abnormality			
Temporal-mandibular joint dysfunction			
Weakness			
Other? Specify:			

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For each indicated issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A frequent feature for this patient

Neoplastic: [] no change from previous entry [] section skipped [] None of the following

	A	B	C
Leukemia			
ALL			
AML			
CLL			
CML			
Unspecified			
Lymphoma- non-Hodgkin's EBV+			
Lymphoma non-Hodgkin's EBV-			
Lymphoma – Hodgkin's EBV+			
Lymphoma – Hodgkin's EBV-			
Polyclonal lymphoproliferation – EBV+			
Polyclonal lymphoproliferation – EBV-			
Solid tumor			
Specify type:			
Monoclonal gammopathy			
Thymoma			
Other?			

Neurologic : [] no change from previous entry [] section skipped [] None of the following

	A	B	C
Ataxia			
Choreoathetosis			
Depression			
Hypotonia			
Intracranial hemorrhage			
Mental retardation			
Microcephaly			
Multiple sclerosis			
Peripheral neuropathy			

	A	B	C
School Failure			
Seizures			
Stroke (CVA)			
Subdural hematoma			
Tetany			
Unexplained neurologic deterioration			
Progressive multifocal leukoencephalopathy (PML)			
Other? Specify:			

Psychosocial: [] no change from previous entry [] section skipped [] None of the following

	A	B	C
ADHD			
Autism			
Behavioral problems			
Depression			
Delinquency			
Held out of school by parents			
Incarceration			
Learning disability			
Psychosis			
School failure			
Other?			

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For each indicated issue, indicate as follows:

A) Present at or before diagnosis B) Observed sometime during patient's illness C) A frequent feature for this patient

Sino-Pulmonary: no change from previous entry section skipped None of the following

	A	B	C
Asthma			
Bronchitis (chronic)			
Bronchiectasis			
Bronchiolitis obliterans			
Bronchiolitis obliterans organizing pneumonia (BOOP)			
Clubbing			

	A	B	C
COPD			
Emphysema			
Granulomas			
Interstitial lung disease			
Pulmonary embolism			
Other? Specify:			

Skin: no change from previous entry section skipped None of the following

	A	B	C
Abnormal hair			
Alopecia			
Anhydrotic ectodermal dysplasia			
Atopic dermatitis/eczema			
Decreased sweating			
Delayed separation of umbilical cord			
Delayed wound healing			
Discoid lupus			
Erythroderma			
Granulomas			
GVHD or GVHD-like rash			
Hypohidrosis			

	A	B	C
Nail defects			
Pruritis			
Psoriasis			
Telangiectasia			
Scarring (abnormal)			
Scleroderma			
Subacute cutaneous lupus			
Sweating (increased)			
Urticaria			
Vasculitis			
Vitiligo			
Other? Specify:			
Other? Specify:			

Allergic Reactions: no change from previous entry section skipped None of the following

If causative agent is known, please indicate corresponding number from table to the right. If other, please specify.

Reaction	A	B	C	Agent (if known)
Anaphylaxis				
Angioedema				
Asthma				
Bronchospasm				
Dermatitis				
Gastrointestinal				
Stevens Johnson				
Urticaria				
Other (Specify):				

List of Agents	
1	Drug
2	Environmental
3	Food
4	Other

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Experience with Live Agent Vaccines List all complications at the end of this section *.

	Vaccine Given			Date Given (mm/dd/yyyy)	Complications		If yes, Describe Outcome
	Yes	No	Unknown		Yes	No	
Polio							
Measles (MMR)							
Varicella							
Vaccinia							
BCG							
Yellow Fever							
FluMist							
Rotavirus							
Other							

*Complications:

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Patient Name, Initials or Site Study Participant #: _____

Treatment History : no change from previous entry section skipped

Immune Globulin therapy: yes , no If yes list starting age ___y___m or date ___/___/___(mm/dd/yyyy)

Current route of administration: iv im sc unknown other (specify): _____

Dose and interval of present therapy:

Dose	Frequency
mg/kg	Every days

Reactions to immunoglobulin treatment?

	Mild	Moderate	Severe
Abdominal pain			
Anaphylaxis			
Anxiety			
Aseptic meningitis			
Fever/Chills			
Headache			
Hepatitis			
Hypertension			
Hypotension			
Local reaction (sc)			
Myalgias			
Renal dysfunction			
Seizure			
Stroke			
Thromboembolism			
Urticaria			
Vomiting			
Wheezing			
Diarrhea			
Severe local reaction			
Other reactions:			

How treated? _____

Has immunoglobulin replacement been stopped because of side effects? yes , no unknown

Explain: _____

Was a port placed specifically for IVIG? Yes No

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Anti-infectives : [] section skipped

Current Anti-infectives

	Drug	Indication	Continuous	Intermittent	Rotating	Duration	Outcome
Antibiotics							
Antibiotics							
Antibiotics							
Antibiotics							
Anti-virals							
Anti-virals							
Anti-fungals (systemic)							
Anti-fungals (systemic)							

Did this treatment lead to significant complications? Yes [], no [], unknown []

*Complications: _____

Immunomodulator therapy : [] section skipped

	Indication	Drug	Duration	Outcome
Non-steroidal anti-inflammatory drugs				
Steroids (systemic)				
Immunosuppressives				
Hydroxychloroquine				
Plasmapheresis				
Rituximab				
TNF alpha inhibitors				
Win-Rho				
Other				
Other				

Did this treatment lead to significant complications? Yes [], no [], unknown []

*Complications: _____

Transfusion treatments : [] section skipped

	Yes	No	How many in past 12 months?	How many in past 5 years?	Irradiated?			
					Yes	No	Both	Unknown
Leukocyte transfusions								
Platelet transfusions								
Red cell transfusions								
Fresh Frozen Plasma								
Whole Blood								

Did this treatment lead to significant complications? Yes [], no [], unknown []

*Complications: _____

Does the patient regularly use non-standard, alternative or complementary treatments?

Yes [] No [] Type? _____

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Surgical and Diagnostic Procedures – please list any complications at the end of this section*

[] no change from previous entry [] section skipped

Surgery (list all complications below)	Yes	No	Age (years)	Date (mm/dd/yyyy)	Response/result
Appendectomy					
Bronchoscopy					
Colonoscopy					
Diagnostic node bx					
Feeding tube					
Gastroscopy					
GI biopsy					
Colon					
Rectum					
Small intestine					
Stomach					
Unspecified					
Indwelling (Central) venous line					
Portacath					
Liver biopsy					
Lung biopsy					
Lung resection					
"PE tubes"					
Sinus surgery					
Splenectomy					
Post splenectomy prophylaxis?					
Antibiotics					
IVIg					
Tonsillectomy					
Other surgeries (list):					

*Complications:

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Transplantation?

Yes [], No [], If yes: Age ___yrs ___mos or date ___/___/_____(mm/dd/yyyy)

If booster or additional transplants used: Age(s) ___/___ (yr./mos.) or date(s) ___/___/_____

If yes: Hematopoietic stem cells [] or solid organ []?

If Hematopoietic stem cells: **(mark all that apply)**

CIBMTR Recipient ID number (CRID) _____

CIBMTR Center number _____

Most significant reason for SCT: Primary immune deficiency [], Malignancy [], Autoimmunity []

Donor: sibling full match [], sibling partial match [], unrelated full match [],

unrelated partial match [], haploidentical – parental donor [], unknown []

Type of graft: mobilized peripheral blood [], cord blood [], bone marrow [], T cell depleted [], unfractionated [], CD34 enriched [], unknown []

Conditioning used: myeloablative [], non-myeloablative [], no conditioning [], unknown []

Was radiation used during conditioning? Yes [] No []

Outcome:

- Engraftment:

Months post transplant	% Donor T cells	% Donor B cells	% Donor myeloid

- Alive with little evidence of engraftment []
- Not engrafted []
- Acute GVHD [] Grade _____
 - Organs involved (list most to least extensively involved)____
- Chronic GVHD [] Grade _____
 - Organs involved____ (list most to least extensively involved)____
- Deceased []; time from transplant to death: _____ [] months or [] years
- Re-transplanted []
- Long term post transplantation immunosuppression required []

Other transplant-related morbidities? Please specify: _____

Comments: _____

Was **gene therapy** used for this patient? Yes [] No []

Specify Gene _____ Specify vector type _____ Which Cells transduced? _____

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If solid organ: **(mark all that apply)**

Which solid organ: kidney [], liver [], lung [], heart [], cornea [], thymus []

Donor: cadaver [], related [], unrelated living [], unknown []

Reason for transplant _____

Outcome:

- stable with good function []
- rejected and re-transplanted []
- stable with acceptable function []
- rejected []
- not stable/ re-transplant probable []
- unknown []

Other? Please specify: _____

Comments: _____

Immunosuppression program? _____

If transplant was considered and not performed, why was transplantation not used?

Solid Organ	
	Lack of a suitably matched donor
	Lack of any donor
	Age or condition of the patient
	Unfavorable probability of success
	Religious objections
	Insurance denial
	Other _____

Hematopoietic Stem Cell	
	Lack of a suitably matched donor
	Lack of any donor
	Age or condition of the patient
	Unfavorable probability of success
	Religious objections
	Insurance denial
	Other _____

Comments: _____

Present Status – Quality of Life

Alive: yes [], no [], unknown []

Date ___/___/_____(mm/dd/yyyy) or age ___yrs ___mos at death

Cause(s) of death? (specify) _____, _____, _____

Does/did patient have significant disability? Full [], partial [], beginning at age ___ / ___ (yr./mos.)

Nature of disability (circle): cognition, vision, hearing, coordination, gait; seizure disorder, pulmonary insufficiency, heart failure, dementia, paralysis, malabsorption/malnutrition, other (specify) _____

Remediation required for missing school? Yes [] No []

Did the patient or parent lose a job as a result of illness? Yes [] No []

Days in hospital during the past 12 months related to PID? _____

Still receiving IVIG? Yes [] No []

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For Adults (>18 years old): Karnofsky index: Mark most appropriate numerical (percentage)

response:

Check here if Karnofsky Index not evaluated	<input type="checkbox"/>
---------------------------------------------	--------------------------

KARNOFSKY PERFORMANCE STATUS SCALE DEFINITIONS RATING (%) CRITERIA

The Karnofsky Performance Scale Index allows patients to be classified as to their functional impairment. This can be used to compare effectiveness of different therapies and to assess the prognosis in individual patients. The lower the Karnofsky score, the worse the survival for most serious illnesses.			Mark here:
Able to carry on normal activity and to work; no special care needed.	100	Normal no complaints; no evidence of disease.	
	90	Able to carry on normal activity; minor signs or symptoms of disease.	
	80	Normal activity with effort; some signs or symptoms of disease.	
Unable to work; able to live at home and care for most personal needs; varying amount of assistance needed.	70	Cares for self; unable to carry on normal activity or to do active work.	
	60	Requires occasional assistance, but is able to care for most of his personal needs.	
	50	Requires considerable assistance and frequent medical care.	
Unable to care for self; requires equivalent of institutional or hospital care; disease may be progressing rapidly.	40	Disabled; requires special care and assistance.	
	30	Severely disabled; hospital admission is indicated although death not imminent.	
	20	Very sick; hospital admission necessary; active supportive treatment necessary.	
	10	Moribund; fatal processes progressing rapidly.	
	0	Dead	

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For Children: Lansky index: Mark most appropriate assessment

Check here if Lansky Index not evaluated	<input type="checkbox"/>
------------------------------------------	--------------------------

- __100 - fully active, normal
- __90 - minor restrictions in strenuous physical activity
- __80 - active, but tires more quickly
- __70 - greater restriction of play *and* less time spent in play activity
- __60 - up and around, but active play minimal; keeps busy by being involved in quieter activities
- __50 - lying around much of the day, but gets dressed; no active playing participates in all quiet play and activities
- __40 - mainly in bed; participates in quiet activities
- __30 - bedbound; needing assistance even for quiet play
- __20 - sleeping often; play entirely limited to very passive activities
- __10 - doesn't play; does not get out of bed
- __0 - unresponsive

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Core Laboratory Test Results

	<u>At or before diagnosis</u>	<u>At current/recent visit</u>
<u>Complete Blood count</u>	(age ___/___ or date ___/___/___) Yr. / Mos. mm/dd/yyyy	(age ___/___ or date ___/___/___) Yr. / Mos. mm/dd/yyyy
WBC	_____ absolute/ul	_____ absolute/ul
Platelets	_____ /ul	_____ /ul
RBC	_____ x 10 ³ /u	_____ x 10 ³ /ul l
Hgb	_____ g/dl	_____ g/dl
Lymphocytes	_____ % _____ /ul	_____ % _____ /ul
PMN	_____ % _____ /ul	_____ % _____ /ul
Eosinophils	_____ % _____ /ul	_____ % _____ /ul
Basophils	_____ % _____ /ul	_____ % _____ /ul
Monocytes	_____ % _____ /ul	_____ % _____ /ul

Lymphocyte Phenotype (age ___/___ (yr./mos.) or date ___/___/___)(mm/dd/yyyy)

Absolute Lymphocyte count	_____ /ul	_____ /ul
CD3 T cells	_____ % _____ /ul	_____ % _____ /ul
CD4 Helper T	_____ % _____ /ul	_____ % _____ /ul
CD8 Cytotoxic T	_____ % _____ /ul	_____ % _____ /ul
CD19 or B cells	_____ % _____ /ul	_____ % _____ /ul
CD20 B cells	_____ % _____ /ul	_____ % _____ /ul
CD56/CD16 NK cells	_____ % _____ /ul	_____ % _____ /ul
CD4+/CD45RA+ (naïve T)	_____ % _____ /ul	_____ % _____ /ul
CD4+/CD45RO+ (memory T)	_____ % _____ /ul	_____ % _____ /ul

Immunoglobulin evaluations (age ___/___ or date ___/___/___) (age ___/___ or date ___/___/___)

Yr./mos. mm/dd/yyyy yr./mos mm/dd/yyyy

<u>At or before diagnosis</u>	<u>At current/recent visit</u>
IgG _____ mg/dl	IgG _____ mg/dl
IgA _____ mg/dl	IgA _____ mg/dl
IgM _____ mg/dl	IgM _____ mg/dl
IgE _____ IU/ml	IgE _____ IU/ml
Yes [] No []	Yes [] No []

Please indicate above if patient received recent Ig replacement or blood products

Antibody Responses (age ___/___) or date ___/___/___ (date closest to diagnosis – before IGIV preferred)

Yr./mos mm/dd/yyyy

	Absent	Low	Normal
Diphtheria	[]	[]	[]
Tetanus	[]	[]	[]
HIB	[]	[]	[]
Protein conjugated HIB	[]	[]	[]
Isohemagglutinin			
Anti-A	[]	[]	[]
Anti-B	[]	[]	[]
Other vaccine antigens			
(specify) _____	[]	[]	[]
(specify) _____	[]	[]	[]
Chicken pox	[]	[]	[]
MMR	[]	[]	[]
ΦX174 or other neoantigen	[]	[]	[]

CORE Dataset: USIDNET Immunodeficiency Patient Registry

Patient Name, Initials or Site Study Participant #: _____

Pneumococcal Polysaccharide After Unconjugated vaccine

Number of serotypes producing a protective level / total serotypes tested from vaccine [/]

After Conjugated Pneumococcal vaccine

Number of serotypes producing a protective level / total serotypes tested from vaccine [/]

	Absent	Low	Normal
unconj PPS serotype _____	[]	[]	[]
unconj PPS serotype _____	[]	[]	[]
unconj PPS serotype _____	[]	[]	[]
Other vaccine _____	[]	[]	[]

Lymphocyte Function (age / (yrs./mos) or date / /) (mm/dd/yyyy)

	Absent	Low	Normal
PHA	[]	[]	[]
ConA	[]	[]	[]
PWM	[]	[]	[]
Anti-CD3	[]	[]	[]
Antigens			
Tetanus	[]	[]	[]
Diphtheria	[]	[]	[]
Candida	[]	[]	[]
Other 1	[]	[]	[]
Other 2	[]	[]	[]
Alloantigen (MLC)	[]	[]	[]
NK cell cytotoxicity	[]	[]	[]

Delayed Hypersensitivity Skin Testing (age ___/___(yrs/mos) or date ___/___/___) (mm/dd/yyyy)

	Absent	Questionable or Weak	Present
Tetanus	[]	[]	[]
Candida	[]	[]	[]
Other 1 _____	[]	[]	[]
Other 2 _____	[]	[]	[]

Complement Function (age ___/___(yrs/mos) or date ___/___/___) (mm/dd/yyyy)

CH50 Absent [] Low [] Normal []

Phagocyte Function (age ___/___(yrs/mos) or date ___/___/___) (mm/dd/yyyy)

Nitro Blue Tetrazolium (NBT) test: normal [] defective []

Neutrophil oxidative burst assay (DHR(dihydrorhodamine)): normal [] defective []

Other test (specify) _____

