Primary Immunodeficiency Diseases: 2017 Clinical Quality Measures

Performance Improvement Module

Developed by:

The Primary Immunodeficiency Diseases Committee of the American Academy of Allergy Asthma and Immunology (AAAAI)

Approved by AAAAI Board of Directors June 2015

Measure Development Workgroup Members

Morna Dorsey, MD MMSc FAAAAI
University of California San Francisco

Ivan Chinn, MD
Baylor College of Medicine

Lisa J. Kobrynski, MD MPH FAAAAI
Emory University

Elena Perez, MD PhD FAAAAI
University of Miami

Panida Sriaroon, MD FAAAAI
University of South Florida

Charlotte Cunningham-Rundles, MD, PhD
The Immunology Institute

Table of Measures	Page

Measure #1: Primary Immunodeficiency: Medical History for Infection – National Quality Strategy Domain: Effective Clinical Care

DESCRIPTION:
Percentage of new patients, regardless of age, with a diagnosis of a primary immunodeficiency for whom an appropriate and thorough medical history regarding infection was used to identify primary immunodeficiency.

INSTRUCTIONS:
The purpose of this measure is to determine if the physician inquired about basic elements of an effective medical history in a patient with suspected primary immunodeficiency. It is not meant to be inclusive of all elements of a pertinent medical history. This measure is to be reported for all new patients seen that have a high index of suspicion for primary immunodeficiency. For the purposes of this measure, pediatric patients will be all patients 0-18 years of age. Adult patients will be all patients 19 years and older. It is anticipated that clinicians who provide the primary management of patients with primary immunodeficiency will report this measure.

DENOMINATOR:
Total number of new patients, regardless of age, with a diagnosis of a primary immunodeficiency or suspected primary immunodeficiency

Denominator Criteria (Eligible Cases):
All new patients regardless of age (pediatric 0-18, adult ≥19 years old)
AND
Diagnosis for a primary immunodeficiency or suspected primary immunodeficiency:

Diagnosis for Immunodeficiency with predominantly antibody defects (ICD-10):
D80.0, D80.1, D80.2, D80.3, D80.4, D80.5, D80.6, D80.7, D80.8, D80.9
Diagnosis for Combined immunodeficiencies (ICD-10):
D81.0, D81.1, D81.2, D81.3, D80.4, D81.5, D81.6, D81.7
Diagnosis for Other combined immunodeficiencies (ICD-10):
D81.810, D81.818, D81.819, D81.89, D81.9
Diagnosis for Immunodeficiency associated with other major defects (ICD-10):
D82.0, D82.1, D82.2, D82.3, D82.4, D82.8, D82.9
Diagnosis for Common variable immunodeficiency (ICD-10):
D83.0, D83.1, D83.2, D83.8, D83.9
Diagnosis for Other immunodeficiencies (ICD-10): D84.0, D84.1, D84.8, D84.9
OR
Patient evaluated for suspected primary immunodeficiency but no PID diagnosis was made

AND
New patient encounter during the reporting period:
New patient office or other outpatient services: 99201, 99202, 99203, 99204, 99205
Initial Hospital Care visit code: 99221, 99222, 99223

Denominator Exclusions: None

NUMERATOR:
Number of patients who were queried about **ALL** of the following during a face-to-face evaluation by a medical professional AND findings were documented in the medical record:

1) Description and frequency of significant lifetime infections (ex: infections requiring hospitalization, IV antibiotics, prolonged antibiotic treatment)
2) Site of infections (ex: pneumonia, osteomyelitis, meningitis)

3) Presence of unusual infections or severe infections (ex: mycobacterial, fungal, opportunistic or disseminated infection)
4) Family history of recurrent infections or primary immunodeficiency

Numerator Options

Performance Met: Patient was queried about ALL of the numerator criteria and findings were documented in the medical record

OR

Performance Not Met: Patient was **not** queried about ALL of the numerator criteria and findings were not documented in the medical record

RATIONALE:

Expert opinion suggests that a careful medical history provides evidence for suspecting primary immunodeficiency and for focusing the evaluation.

The following rationale is copied verbatim from the listed clinical guideline:

Summary Statement 1:
PIDD usually presents with signs and symptoms of infections that may be repetitive, severe, or refractory to therapy and caused by organisms of low virulence. Infection is by far the most common complication of PIDD and the most frequent problem that leads to medical evaluation. Infections in immunodeficient patients usually occur with pathogens that are prevalent in the community but are of unusual severity, frequency, and duration. They also tend to respond poorly to therapy. Children with invasive pneumococcal disease should undergo immunologic investigation, as up to 26% of these patients older than 2 years have an identifiable primary immunodeficiency. Severe PIDD such as severe combined immunodeficiency (SCID) and many others may also be associated with infections caused by low-grade or opportunistic organisms that are rarely pathogenic for immunocompetent individuals.

Summary statement 4. A focused family history (e.g. recurrent infections, absence of infections in siblings, early childhood deaths, diagnosed PIDD) should be obtained when the differential diagnosis includes PIDD. (D)

Supporting Guideline: Practice parameter for the diagnosis and management of primary immunodeficiency. Bonilla, FA et al. The Journal of Allergy Clinical Immunology 2015,136(5):1186-205,e1-78..

Measure Type: Process

Measure #2: Antibody Deficiency Disorder: Appropriate Laboratory Testing – National Quality Strategy Domain: Effective Clinical Care

DESCRIPTION:
Percentage of new patients aged 2 years and older with a suspected antibody deficiency disorder who had an appropriate work up for antibody deficiency disorders including laboratory testing for IgG level, IgA level, IgM level, protein IgG antibody response and polysaccharide IgG antibody response.

INSTRUCTIONS:
This measure is to be reported on patients with a suspected antibody deficiency disorder. The purpose of this measure is to determine if the physician obtained basic elements of an initial evaluation for a patient with suspected antibody deficiency. It determines if an appropriate work up for antibody deficiency disorders was performed by confirming that five laboratory tests were ordered and reviewed prior to making a formal diagnosis. These studies include laboratory testing for total serum IgG, IgA, IgM levels, protein IgG antibody responses, and polysaccharide IgG antibody responses. All 5 tests must be reviewed, *using age appropriate normal values,* in order to meet the measure. This measure is not intended to be inclusive of all elements of a pertinent laboratory investigation (such as IgG subclasses, isohemagglutinin titers, lymphocyte subsets) or to be sufficient for prescribing treatment. *Measurement of specific IgG to a protein (e.g. tetanus, diptheria toxoid) or a polysaccharide (e.g. pneumococcus, meningococcus) antigen should be measured in immunized patients.* Other tests that measure these elements, such as antibody response to immunization with neo-antigens bacteriophage **ϕX174 or** keyhole limpet hemocyanin **(KLH)** are currently only available on investigator IND and conducted on a research basis. The pediatric population is limited to 2-18 years of age. Adult patients will be all patients 19 years and older. It is anticipated that clinicians who provide the primary management of patients with primary immunodeficiency will report this measure.

DENOMINATOR:
Total number of new patients, aged 2 years and older, with a suspected antibody deficiency disorder.

Denominator Criteria (Eligible Cases):
New patients aged 2 years and older
AND
Diagnosis of antibody deficiency disorder or clinical suspicion for an antibody deficiency disorder:
Diagnosis for Immunodeficiency with predominantly antibody defects (ICD-10):
D80.0, D80.1, D80.2, D80.3, D80.4, D80.5, D80.6, D80.7, D80.8, D80.9
OR
Patient evaluated for suspected antibody deficiency disorder but diagnosis for an antibody deficiency disorder was not made
AND
New patient encounter during the reporting period:
New patient office or other outpatient services: 99201, 99202, 99203, 99204, 99205
Initial Observational Care, New or Established Patient: 99218, 99219, 99220
Initial Hospital Care: 99221, 99222, 99223

Denominator Exclusions: Patient has a previously diagnosed primary immunodeficiency or is receiving immunoglobulin (IgG therapy)

NUMERATOR:
Number of patients who had **ALL** of the following laboratory investigations with results documented in the medical record:
1) Total serum IgG level
2) Total serum IgA level
3) Total serum IgM level
4) Protein IgG antibody response (e.g. Tetanus toxoid, Diphtheria toxoid, H. influenza)

5) Polysaccharide IgG antibody response (e.g. Streptococcus pneumoniae PPSV-23)

Numerator Options

Performance Met: Laboratory testing for total serum IgG level, IgA level, IgM level, protein IgG antibody response and polysaccharide IgG antibody response were reviewed and results were documented in the medical record

OR

Performance Not Met: Laboratory testing for total serum IgG level, IgA level, IgM level, protein IgG antibody response and polysaccharide IgG antibody response were **not** reviewed and results were not documented in the medical record

OR

Other Performance Exclusion: Clinician has documented reason for not performing laboratory testing (eg, patient declined, other patient reasons, absent or very low IgG levels which would not necessitate vaccine or antibody levels, patient has never received vaccine)

RATIONALE:

Expert opinion suggests that important laboratory investigations be conducted in the work up of antibody deficiency disorders.

The following rationale is copied verbatim from the listed clinical guideline:

Summary Statement 3: There are 4 primary immunodeficiencies that largely depend on qualitative analysis of vaccination responses. (IV D)

Immunologists rely on the vaccine response to help make the distinction between significant PIDDs and transiently low immunoglobulin levels, as seen in patients with THI, or delayed maturation of antibody responsiveness. If the vaccine response is interpreted as normal, a diagnosis of some form of delayed maturation of antibody responsiveness might be likely, common humoral immunodeficiencies (ie, selective IgA deficiency with IgG subclass deficiency and selective antibody deficiency) rely on accurate interpretation of vaccine responses for appropriate diagnosis and management.

Supporting Guideline: Use and interpretation of diagnostic vaccination in primary immunodeficiency: A working group report of the Basic and Clinical Immunology Interest Section of the American Academy of Allergy, Asthma & Immunology. Orange, JS et al. Journal of Allergy Clinical Immunology 2012;130:S1-24.

Summary statement 6. Evaluation of specific immune responses is essential for diagnosis of PIDD. (C)
Measurement of serum immunoglobulin levels and lymphocyte responses to mitogens are useful indicators of global B-cell and T-cell development and function. However, these studies may appear normal in many primary immunodeficiencies, because they are not sensitive indicators of specific immunity: the responses of T and B cells to antigen. For evaluation of humoral immune function, specific antibody titers to both protein and polysaccharide antigens should be measured. These substances differ in how they stimulate antibody production, and clinically significant disease may result from a selective inability to respond to polysaccharide antigens (see SS 105). Note that in individuals with findings consistent with agammaglobulinemia (see section on antibody deficiencies) measurement of specific antibody responses may not be necessary.

Supporting Guideline: Practice parameter for the diagnosis and management of primary immunodeficiency. Bonilla, FA et al. The Journal of Allergy Clinical Immunology 2015, 136(5):1185-205.

Measure Type: Process

Measure #3: Primary Immunodeficiency: Vaccination Recommendation for Close Contacts – National Quality Strategy Domain: Communication and Care Coordination

DESCRIPTION:
Percentage of patients, regardless of age, with a diagnosed primary immunodeficiency requiring IgG therapy, who received counseling regarding the importance of immunizations in those in close contact with them.

INSTRUCTIONS:
This measure is to be reported for contacts of patients with a primary immunodeficiency disease diagnosis requiring IgG therapy. The intent of this measure is to determine if the physician documented a discussion regarding herd immunity by recommending the patient's close contacts stay up-to-date on CDC vaccine recommendations such as annual influenza vaccine and other scheduled vaccines. Further, the clinician should discuss and document the risks, benefits and possible exceptions of vaccinations. This measure is to provide vaccine recommendations for close patient contacts and not for the patient themselves. For the purposes of this measure, the pediatric population is 0-18 years of age. Adult patients will be all patients 19 years and older. It is anticipated that clinicians who provide the primary management of patients with primary immunodeficiency will report this measure.

DENOMINATOR:
Number of patients, regardless of age, with a diagnosed primary immunodeficiency.

Denominator Criteria (Eligible Cases):
All new patients regardless of age (pediatric 0-18, adult ≥19 years old)
AND
Diagnosis for a primary immunodeficiency

Diagnosis for Immunodeficiency with predominantly antibody defects (ICD-10):
D80.0, D80.1, D80.2, D80.3, D80.4, D80.5, D80.6, D80.7, D80.8, D80.9
Diagnosis for Combined immunodeficiencies (ICD-10):
D81.0, D81.1, D81.2, D81.3, D81.5, D81.6, D81.7
Diagnosis for Other combined immunodeficiencies (ICD-10):
D81.810, D81.818, D81.819, D81.89, D81.9
Diagnosis for Immunodeficiency associated with other major defects (ICD-10):
D82.0, D82.1, D82.2, D82.3, D82.4, D82.8, D82.9
Diagnosis for Common variable immunodeficiency (ICD-10):
D83.0, D83.1, D83.2, D83.8, D83.9
Diagnosis for Other immunodeficiencies (ICD-10): D84.0, D84.1, D84.8, D84.9

AND
Patient receiving IgG therapy
96365, 96366, 96372

AND
Patient encounter during the reporting period: 99201, 99202, 99203, 99204, 99205, 99211, 99212, 99213, 99214, 99215

Denominator Exclusions: None

NUMERATOR:
The number of patients with documented verbal or written recommendations regarding immunizations with inactivated influenza vaccine or other immunizations for close patient contacts AND discussion of risks, benefits and exceptions.

Numerator Note: For the purposes of this measure possible immunizations include but are not limited to influenza vaccine, pertussis vaccine (Tdap), MMR (measles, mumps, rubella) vaccine, varicella vaccine (for older contacts if not immune)

Numerator Options

Performance Met: Documented verbal or written recommendations regarding immunizations with inactivated influenza vaccine or other immunizations for close patient contacts with discussion of risks, benefits and exceptions.

OR

Performance Not Met: **No** documented verbal or written recommendations regarding immunizations with inactivated influenza vaccine or other immunizations for close patient contacts with discussion of risks, benefits and exceptions

RATIONALE:

Expert opinion suggests that close contacts be vaccinated to prevent infection in the immunocompromised host. Annual immunizations with inactivated influenza vaccine is recommended.

The following rationale is copied verbatim from the listed publication by the Medical Advisory Committee of the Immune Deficiency Foundation:

Close contacts of patients with compromised immunity should not receive live oral poliovirus vaccine because they might shed the virus and infect a patient with compromised immunity. Close contacts can receive other standard vaccines such as Measles, Mumps, Rubella, Varicella (MMRV), pertussis (Tdap) and hepatitis because viral shedding is unlikely and these pose little risk of infection to a subject with compromised immunity. Particularly important are annual immunizations with inactivated influenza vaccine; scheduled periodic pertussis vaccine; pneumococcal vaccine; measles, mumps, and rubella vaccine; and varicella vaccine for older contacts whose routine immunizations might not be up to date.

Supporting Guideline: Recommendations for live viral and bacterial vaccines in immunodeficient patients and their close contacts. Medical Advisory Committee of the Immune Deficiency Foundation. Shearer, WT et al. Journal of Allergy and Clinical Immunology. 2014 Apr;133(4):961-6.

Measure Type: Process

Measure #4: Primary Immunodeficiency: IgG Therapy – National Quality Strategy Domain: Effective Clinical Care

DESCRIPTION:
Percentage of patients, regardless of age, with a diagnosed primary immunodeficiency requiring IgG therapy who were assessed prior to IgG dose or interval adjustment or for the purposes of monitoring patient outcomes.

INSTRUCTIONS:
This measure is to be reported on patients with a diagnosed primary immunodeficiency requiring IgG therapy. Any changes in IgG therapy should be made with careful and due consideration by the clinician. The intent of this measure is to determine if the physician documented basic medical history to inform appropriate dosing and method of delivery of immunoglobulin replacement product. It does not exclude other pertinent elements of the history. It is also not designed to address monitoring for other complications such as disease transmission, hemolysis or kidney function. In order to fulfill this measure, the clinician must query the patient for the number of infections since the last visit, asses the type of infections since the last visit, reactions during or after infusions and obtain a serum trough level (or steady-state if on subcutaneous IgG) at least once in the last 12 months. Clinical rationale should be documented taking into account this information and the patient's individual characteristics, prior to dosing and interval adjustments or for the purpose of monitoring outcomes. For the purposes of this measure, the pediatric population is 0-18 years of age. Adult patients will be all patients 19 years and older. It is anticipated that clinicians who provide the primary management of patients with primary immunodeficiency will report this measure.

DENOMINATOR:
Number of patients, regardless of age, with a diagnosed primary immunodeficiency requiring IgG therapy

Denominator Criteria (Eligible Cases):
All patients regardless of age (pediatric 0-18, adult ≥ 19 years old)
AND
Diagnosis with a primary immunodeficiency

Diagnosis for Immunodeficiency with predominantly antibody defects (ICD-10):
D80.0, D80.1, D80.2, D80.3, D80.4, D80.5, D80.6, D80.7, D80.8, D80.9
Diagnosis for Combined immunodeficiencies (ICD-10):
D81.0, D81.1, D81.2, D81.3, D81.5, D81.6, D81.7
Diagnosis for Other combined immunodeficiencies (ICD-10):
D81.810, D81.818, D81.819, D81.89, D81.9
Diagnosis for Immunodeficiency associated with other major defects (ICD-10):
D82.0, D82.1, D82.2, D82.3, D82.4, D82.8, D82.9
Diagnosis for Common variable immunodeficiency (ICD-10):
D83.0, D83.1, D83.2, D83.8, D83.9
Diagnosis for Other immunodeficiencies (ICD-10): D84.0, D84.1, D84.8, D84.9

AND
Patient receiving IgG therapy
96365, 96366, 96372

AND
Patient encounter during the reporting period: 99201, 99202, 99203, 99204, 99205, 99211, 99212, 99213, 99214, 99215

Denominator Exclusions: None

NUMERATOR:
Number of patients who were queried and assessed for **ALL** of the following prior to an adjustment in IgG replacement therapy or for the purpose of monitoring patient outcomes:

1) Number of infections since the last visit
2) Type of infections since the last visit
3) Reactions during or after infusions
4) Serum trough (or steady-state if on subcutaneous IgG) at least once in the last 12 months

Numerator Options

Performance Met:	Patient was queried and assessed for ALL of the numerator criteria since the last visit
OR	
Performance Not Met:	Patient was **not** queried and assessed for ALL of the numerator criteria since the last visit

RATIONALE:
Expert opinion suggests that practitioners be familiar with the biological trough level and that this should be the serum IgG trough level that best improves the patient's clinical course and diminishes infections. Practitioners should be aware of the importance of monitoring clinical outcomes, including serum IgG levels while on therapy.

The following rationale is copied verbatim from the listed publication:

Guiding Principal 5: IgG trough levels - IgG trough levels can be useful in some diagnoses to guide care but are NOT useful in many and should NOT be a consideration in access to IVIG therapy.

Supporting Guideline: Eight Guiding Principles for Effective Use of IVIG for Patients with Primary Immunodeficiency 2011. Update on Use of Immune Globulin (IG) in Human Disease: A review of evidence by members of the primary immunodeficiency committee of the American Academy of Allergy, Asthma and Immunology. 2015 Perez, EE.

The following rational is included with permission from Dr. Elena Perez from her article currently under review entitled: *"Update on Use of Immune Globulin (IG) in Human Disease: A review of evidence by members of the primary immunodeficiency committee of the American Academy of Allergy, Asthma and Immunology"*

"An acceptable starting point for maintenance dosing is 400 to 600 mg mg/kg every three to four weeks and is consistent with majority practice by focused US and European immunologists.[550, 551] Trough IgG levels do not need to be measured frequently; annually is often satisfactory, however physicians should be aware of weight changes in growing children and adjust dose accordingly. They should be obtained whenever a significant infection occurs or when the clinical response to treatment does not meet expectations. After the fifth infusion, a steady state will have been achieved, and the dose or dosing interval should be adjusted to achieve the optimal clinical result. Periodic [PE1] measurement of trough IgG levels may detect non-compliance by patients who are receiving infusions with home care (or self-administering SCIG at home). "

The workgroup will remove this reference if the article has not been accepted for publication by the completion of the PID practice improvement module.

Measure Type: Intermediary Outcome

Measure #5: Common Variable Immune Deficiency: Pulmonary Care – National Quality Strategy Domain: Person and Caregiver-Centered Experience and Outcomes

DESCRIPTION:
Percentage of patients aged 4 years and older with a diagnosis of common variable immune deficiency (CVID) in which pulmonary disease was considered.

INSTRUCTIONS:
The purpose of this measure is to ensure discussion between the immunologist and patient regarding potential pulmonary complications in both symptomatic and asymptomatic patients. In order to meet this measure, at minimum there should be documentation that a discussion with the patient took place regarding a pulmonary care plan taking into account the patient's age, medical history, radiation exposure, laboratory investigations, review of systems and other factors as appropriate to identify pulmonary complications. This measure is to be reported on patients with diagnosed common variable immune deficiency (CVID). For the purposes of this measure, the pediatric population is 4-18 years of age. Adult patients are all patients age 19 years and older. It is anticipated that clinicians who provide the primary management of patients with primary immunodeficiency will report this measure.

DENOMINATOR:
Number of established patients aged 4 years and older with a diagnosis of common variable immunodeficiency

Denominator Criteria (Eligible Cases):
All patients regardless of age (pediatric 4-18, adult ≥19 years old)
AND
Diagnosis of common variable immunodeficiency (ICD-10): D83.0, D83.1, D83.2, D83.8, D83.9
AND
Patient encounter during the reporting period (CPT): 99201, 99202, 99203, 99204, 99205, 99211, 99212, 99213, 99214, 99215

Denominator Exclusions: None

NUMERATOR:
Number of patients for whom there is a documented discussion regarding pulmonary disease AND an appropriate plan was developed based on individual patient assessment

Numerator Options

Performance Met:	Documentation of discussion regarding pulmonary disease and pulmonary plan of care
OR	
Performance Not Met:	No documentation of discussion regarding pulmonary disease or pulmonary plan of care

RATIONALE:
Expert opinion suggests that presence or absence of lung disease be identified at diagnosis and if present followed closely.

The following rationale is copied verbatim from the listed publication:

Although there is no consensus for monitoring lung damage in CVID patients, pulmonary lung function tests (carbon monoxide diffusion) and high-resolution computed tomography are the best recommended examinations for patients with pulmonary disease. While chest X-rays are not as revealing as HRCT, given the concern for excessive radiation exposure over time it is reasonable to obtain chest X-ray in asymptomatic patients at baseline. Full pulmonary lung function tests should be conducted at yearly

intervals in patients with pulmonary disease. For asymptomatic patients yearly spirometry should be conducted.

Supporting Guideline: Clinical Management Review: The Variable in Common Variable Immunodeficiency. Stephen Jolles. Journal of Allergy and Clinical Immunology in Practice November-December, 2013 Volume 1, Issue 6, Pages 545-556.

Chest x-rays are not as revealing as HRCT, so it is reasonable to obtain these at baseline referral. However, radiosensitivity has been demonstrated in CVID and for a younger subject, yearly or examinations every 2 years, especially in concert with other x-ray procedures, could lead to excessive radiation exposure over time. For more frequent follow-up of patients with chronic cough and/or known lung damage, I prefer complete lung functions, including carbon monoxide diffusion as a means of assessing lung damage at shorter intervals, with possible HRCT at 3- to 4-year intervals or at less frequent intervals to monitor changes in therapy.

Cunningham-Rundles C. How I treat common variable immunodeficiency. Blood (2010) 116:7–15. doi:10.1182/blood-2010-01-254417

Measure Type: Process

Appendix 1: ICD-10 Codes

	Immunodeficiency with predominantly antibody defects
D80.0	Hereditary hypogammaglobulinemia
D80.1	Nonfamilial hypogammaglobulinemia
D80.2	Selective deficiency of immunoglobulin A [IgA]
D80.3	Selective deficiency of immunoglobulin G [IgG] subclasses
D80.4	Selective deficiency of immunoglobulin M [IgM]
D80.5	Immunodeficiency with increased immunoglobulin M [IgM]
D80.6	Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
D80.7	Transient hypogammaglobulinemia of infancy
D80.8	Other immunodeficiencies with predominantly antibody defects
D80.9	Immunodeficiency with predominantly antibody defects, unspecified
	Combined immunodeficiencies
D81.0	Severe combined immunodeficiency [SCID] with reticular dysgenesis
D81.1	Severe combined immunodeficiency [SCID] with low T- and B-cell numbers
D81.2	Severe combined immunodeficiency [SCID] with low or normal B-cell numbers
D81.3	Adenosine deaminase [ADA] deficiency
D81.4	Nezelof's syndrome
D81.5	Purine nucleoside phosphorylase [PNP] deficiency
D81.6	Major histocompatibility complex class I deficiency
D81.7	Major histocompatibility complex class II deficiency
	Other combined immunodeficiencies
	Biotin-dependent carboxylase deficiency
D81.810	Biotinidase deficiency
D81.818	Other biotin-dependent carboxylase deficiency
D81.819	Biotin-dependent carboxylase deficiency, unspecified
D81.89	Other combined immunodeficiencies
D81.9	Combined immunodeficiency, unspecified
	Immunodeficiency associated with other major defects
D82.0	Wiskott-Aldrich syndrome
D82.1	Di George's syndrome
D82.2	Immunodeficiency with short-limbed stature
D82.3	Immunodeficiency following hereditary defective response to Epstein-Barr virus
D82.4	Hyperimmunoglobulin E [IgE] syndrome
D82.8	Immunodeficiency associated with other specified major defects
D82.9	Immunodeficiency associated with major defect, unspecified
	Common variable immunodeficiency
D83.0	Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
D83.1	Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
D83.2	Common variable immunodeficiency with autoantibodies to B- or T-cells
D83.8	Other common variable immunodeficiencies
D83.9	Common variable immunodeficiency, unspecified
	Other immunodeficiencies
D84.0	Lymphocyte function antigen-1 [LFA-1] defect

D84.1	Defects in the complement system
D84.8	Other specified immunodeficiencies
D84.9	Immunodeficiency, unspecified
Z13.0	Encounter for screening for diseases of the blood and blood-forming organs and certain disorders involving the immune mechanism
Z13.9	Encounter for screening, unspecified

Ref: http://www.icd10data.com/ICD10CM/Codes

Appendix 2: Denominator Qualification Questions from PID PIM

Chart Audit Questions:

Measure 1:
(User must respond with YES to all of the questions in order to qualify for the measure)
Chart Audit questions:
During a face-to-face evaluation in order to identify primary immunodeficiency was the patient queried about the following:

Question 1: Description and frequency of significant lifetime infections (ex: infections requiring hospitalization, IV antibiotics, prolonged antibiotic treatment)
Responses:
Yes
No

Question 2: Site of infections (ex: pneumonia, osteomyelitis, meningitis)
Responses:
Yes
No

Question 3: Presence of unusual infections or severe infections (ex: mycobacterial, fungal, opportunistic or disseminated infection)
Responses:
Yes
No

Question 4: Family history of recurrent infections or primary immunodeficiency
Responses:
Yes
No

Question 5: Were all findings were documented in the medical record:
Responses:
Yes
No

Measure 2:

1. Was laboratory testing for total serum IgG level, IgA level, IgM level, protein IgG antibody response and polysaccharide IgG antibody response reviewed?
   1.1. Total serum IgG level
   Responses:
   Yes
   No

1.2. Total serum IgA level
Responses:
Yes
No

1.3. Total serum IgM level
Responses:
Yes
No

1.4. Protein IgG antibody response (ex. Tetanus toxoid, diphtheria toxoid)
Responses:
Yes
No

1.5. Polysaccharide IgG antibody response (ex. Streptococcus pneumonia PPSV-23)
Responses:
Yes
No

2. Were results documented in the medical record?

Responses:
Yes
No

Measure 3:

1. Was there documented verbal or written recommendations regarding immunizations with inactivated influenza vaccine or other immunizations for close patient contacts?
   1.1. Verbal recommendations
   1.2. Written recommendations
   1.3. No recommendations given
2. Was there discussion of risks, benefits and exceptions?
   Responses:
   Yes
   No

Measure 4:

1. Was the patient queried and assessed for **ALL** of the following prior to an adjustment in IgG replacement therapy or for the purpose of monitoring patient outcomes:
   1.1. Number of infections since the last visit
   Responses:
   Yes
   No

   1.2. Type of infections since the last visit
   Responses:
   Yes
   No

   1.3. Reactions during or after infusions

Responses:
Yes
No

1.4. Serum trough (or steady-state if on subcutaneous IgG) at least once in the last 12 months
Responses:
Yes
No

Measure 5:

1. Was there a documented discussion regarding pulmonary disease?
   Responses:
   Yes
   No

2. Was an appropriate pulmonary plan of care developed based on individual patient assessment?
   Responses:
   Yes
   No