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CLINICAL DOCUMENTATION

# Primary Immunodeficiency (IEI)

Immunodeficiency refers to a weakened or dysfunctional immune system, making individuals more susceptible to infections. This can be caused by genetic defects (primary immunodeficiency) or by external factors like illnesses, medications, or nutritional deficiencies (secondary immunodeficiency). Inborn errors of immunity (IEI; previously "primary immunodeficiency diseases") encompass over 485 intrinsic defects of immunity, most of which are inherited.

## ICD-10 CODES

**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 w/ 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 (Kappa light chain deficiency)
**D80.9** Immunodeficiency with predominantly antibody defects, unspecified
**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
**D81.30** Adenosine deaminase deficiency, unspecified
**D81.31** Severe combined immunodeficiency due to adenosine deaminase deficiency
**D81.32** Adenosine deaminase 2 deficiency
**D81.39** Other adenosine deaminase 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
**D81.810** Biotinidase deficiency
**D81.818** Other biotin-dependent carboxylase deficiency
**D81.819** Biotin-dependent carboxylase deficiency, unspecified
**D81.82** Activated Phosphoinositide 3-kinase Delta Syndrome [APDS]
**D81.89** Other combined immunodeficiencies
**D81.9** Combined immunodeficiency, unspecified
**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
**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

## DOCUMENTATION ACRONYMS

### DEEP Diagnosis Elements

Include elements of DEEP in documentation to clinically support primary immunodeficiency.

**Diagnosis:** Immunodeficiency

**Evidence:** T-cell deficiency noted on abs, FISH assay positive for deletion of 22q11.2

**Evaluation:** Di George's syndrome, attributing to hypoparathroidism

**Plan:** Referral to cardiology and endocrinology, continue with calcium and vit D supplementation for hypoparathyroidism

### Final Assessment Details

Include DSP for each addressed condition impacting treatment and patient care.

**Diagnosis:**

**Primary Immunodeficiency Diagnosis**

- Immunodeficiency Type
- Cause

**Status:**

**Active**

- Current symptoms
- Control status

**Plan:**

- Pharmacologic
- Control of symptoms
- Referrals
- Lifestyle changes
- Monitoring for infections

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## BEST PRACTICES & TIPS

- **Specificity is key!** Always indicate the type of IEI, the genetic cause, and use verbiage to represent any complications derived from the defect.
- The use of the term immunosuppressed is is **not synonymous** with a primary immunodeficiency or inborn immunosuppression and should be avoided.
- When documenting immunodeficiency and its etiology, be sure to document **all compounding confirmed factors** to get a complete picture of the patients' health status.
- DSP should be applied for all diseases **as well as** for the resulting immunodeficiency. Status should be apparent by using descriptive words to clarify the presence and severity of the illnesses. (Symptomatic, controlled, uncontrolled, etc.)
- Documentation should **always include DEEP elements** to show evidence for any IEI by incorporating labs, imaging, signs and symptoms and documenting any associated treatments with the corresponding final diagnosis.
- **Avoid** documenting primary immunodeficiencies as a "history of" as this suggests a resolved status and causes conflict within the documentation.
- Confirmation should be found within the documentation representing the **cause and effect** relationship between any infection that is attributed to the presence of an IEI.

For more resources go to:

HIOSCAR.COM/PROVIDERS/RESOURCES