

## Disorder of Immunity

At Healthfirst, we are committed to helping providers accurately document and code their patients' health records. This tip sheet is intended to assist providers and coding staff with the documentation and ICD-10-CM selection, on services submitted to Healthfirst—specifically for **Disorder of Immunity**. It provides information from industry sources about proper coding practice. However, this document does not represent or guarantee that Healthfirst will cover and/or pay for the services outlined. Coverage decisions are based on the terms of the applicable evidence of coverage and the provider's participation agreement. This includes the determination of any amount that Healthfirst or the member owes the provider.

**Disorders of immunity** can be broadly classified into two main categories: Immunodeficiency Disorders and Autoimmune Disease.

### Diagnosis Codes and Descriptions

ICD-10-CM	Description
D70.0	Congenital agranulocytosis
D70.4	Cyclic neutropenia
D71	Functional disorders of polymorphonuclear neutrophils
D72.0	Genetic anomalies of leukocytes
D76.1	Hemophagocytic lymphohistiocytosis
D76.3	Other histiocytosis syndromes
D80.0	Hereditary hypogammaglobulinemia
D80.2	Selective deficiency of immunoglobulin A [IgA]

# Documentation and Coding

ICD-10-CM	Description
<b>D80.3</b>	Selective deficiency of immunoglobulin G [IgG] subclasses
<b>D80.4</b>	Selective deficiency of immunoglobulin M [IgM]
<b>D80.5</b>	Immunodeficiency with increased immunoglobulin M [IgM]
<b>D81.4</b>	Nezelof's syndrome
<b>D82.0</b>	Wiskott-Aldrich syndrome
<b>D82.1</b>	Di George's syndrome
<b>D84.1</b>	Defects in the complement system
<b>D89.84</b>	IgG4-related disease

## Coding Tips

- Screening for blood diseases and/or immune disorders use code **Z13.0**
- When patients are genetically susceptible use code **Z15.89**
- Assign the following codes to specify **past history**:
  - **Z86.2** - Personal history of blood and other disorders involving the immune mechanism
  - **Z83.2** - Family history of blood and other disorders involving the immune mechanism
- Immunodeficiency is associated with other major defects:
  - Wiskott-Aldrich Syndrome (**D82.0**)
  - DiGeorge's Syndrome (**D82.1**)
  - Immunodeficiency with short-limbed stature
  - Immunodeficiency following hereditary defective response to Epstein-Barr virus
  - Hyperimmunoglobulin E (Ig E) Syndrome

# Documentation and Coding

## Clinical Documentation Should Include:

<b>Updated Status of Condition</b>	<ul style="list-style-type: none"><li>• Stable</li><li>• Improved</li><li>• Worsening</li></ul>
<b>Type of Immunity</b>	<ul style="list-style-type: none"><li>• Innate</li><li>• Adaptive</li><li>• Passive</li><li>• Antibody deficiency</li><li>• Cell-mediated</li></ul>
<b>Risk Factors</b>	<ul style="list-style-type: none"><li>• Age</li><li>• Medication (ex: Chemotherapy)</li><li>• Environmental factors</li><li>• Genetics/ Family History</li><li>• Malnutrition</li></ul>
<b>Underlying Causes</b>	<ul style="list-style-type: none"><li>• Genetic disorders</li><li>• Infections (ex: AIDS)</li><li>• Certain Cancers</li><li>• Diabetes</li></ul>
<b>Treatment Plan</b>	<ul style="list-style-type: none"><li>• Immunosuppressants/ Corticosteroids</li><li>• Disease-modifying antirheumatic drugs</li><li>• Stem Cell Transplantation</li><li>• Biologics</li><li>• Physical Therapy</li><li>• Surgery</li><li>• Enzyme Replacement Therapy</li><li>• Gene Therapy</li></ul>

# Documentation and Coding

## Coding Example

<b>Case</b>	A patient with known hereditary hypogammaglobulinemia, refractory multiple myeloma, and recent stem cell transplant was admitted with fever and diarrhea. Further examination revealed tachycardia, leukocytosis, and hypotension. The provider diagnosed hereditary hypogammaglobulinemia, and toxic encephalopathy due to stem cell transplant. What are the appropriate diagnosis code assignments for the admission?
<b>ICD-10 Code and Rationale</b>	Assign Code <b>T86.5</b> - Complications of stem cell transplant, as principal diagnosis for the stem cell transplant complication. Assign codes <b>D80.0</b> , Hereditary hypogammaglobulinemia, and <b>G92</b> , Toxic encephalopathy. Code <b>C90.00</b> Multiple myeloma not having achieved remission, is assigned for the multiple myeloma.

## References

- [AHA Coding Clinic](#)
- [ICD-10-CM Official Coding Guidelines FY 2025](#)
- [Overview of Immunodeficiency Disorders | Merck Manual](#)
- [Primary Immunodeficiency | Mayo Clinic](#)

## Questions?

Contact us at [#Risk\\_Adjustments\\_and\\_clinical\\_Documentation@healthfirst.org](mailto:#Risk_Adjustments_and_clinical_Documentation@healthfirst.org).

For additional documentation and coding guidance, please visit the coding section on [HFproviders.org](https://www.healthfirst.org/HFproviders.org)

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