AAVBC
AMERICAN ACADEMY OF VALUE BASED CARE

Pemphigus

Quick Reference Guide

2026

Table of Contents

1 CLINICAL SNAPSHOT

Definition: Pemphigus is a group of **rare**, potentially life-threatening chronic autoimmune blistering disorders in which **IgG autoantibodies target desmoglein 1 (Dsg[1]) and/or desmoglein 3 (Dsg[3])** - the desmosomal proteins that anchor keratinocytes to one another.[1] Antibody binding triggers acantholysis (loss of cell-to-cell adhesion)[1], producing fragile, flaccid blisters that rupture into **painful, slow-healing erosions on skin and mucous membranes**.[1] **Pemphigus vulgaris** (PV) is the most common subtype and characteristically begins in the oral cavity, often several months before skin involvement appears.[1] Anti-Dsg[1] and/or anti-Dsg[3] IgG are detectable by ELISA in roughly **98.5%** of pemphigus sera.[2] In older adults the leading causes of death are not the blisters themselves but treatment-related complications - pneumonia, septicemia, and cardiovascular events - driven largely by high-dose corticosteroid exposure. First-line rituximab markedly reduces this treatment-related harm across cardiovascular and metabolic outcomes.[2,3]

ICD-10 Codes:

Pemphigus is reported with subtype-specific codes in the bullous disorders category. **L10.0** (pemphigus vulgaris), **L10.1** (pemphigus vegetans), **L10.2** (pemphigus foliaceus), **L10.3** (Brazilian pemphigus/fogo selvagem), **L10.4** (pemphigus erythematosus), **L10.5** (vaccine or drug-induced — also code the causative agent with the appropriate T36-T50 adverse-effect code), **L10.81** (paraneoplastic — also code the associated neoplasm), and **L10.89** (other pemphigus, including IgA pemphigus) all map to **HCC 387**. **L10.9** (pemphigus, unspecified) should be upgraded to the confirmed subtype once direct immunofluorescence (DIF) and ELISA results are available. Commonly co-occurring conditions are coded separately: **E44.0**(moderate protein-calorie malnutrition), **E86.0** (dehydration), **E09.65** (drug-induced diabetes — use **E09.x**, not **E11.x**), **M81.8** with **Z79.52** (drug-induced osteoporosis), **G72.0** (steroid-induced myopathy), **Z79.52** (long-term systemic steroids), and **Z79.620** (long-term immunomodulator/biologic such as rituximab).[4]

Prevalence and Burden: Pemphigus is rare, with US estimates of roughly **5.2 per 100,000** in the general population with prevalence in populations of Ashkenazi Jewish ancestry.[5] The condition disproportionately affects older adults - the median age at pemphigus vulgaris presentation is about **71 years** in population-based studies - making it highly relevant to Medicare Advantage panels.[6] Age is the strongest independent predictor of mortality: patients aged 65 or older at diagnosis face an adjusted hazard ratio of **5.9 for 5-year** and **4.2 for 10-year mortality** compared with younger patients; hypoalbuminemia (<3.5 g/dL) independently predicts 10-year mortality **(HR 3.3)**.[3] Among hospitalized patients, over **50%** carry a serious infection diagnosis, and pneumonia and septicemia are the leading causes of inpatient death.[7]

HCC/RAF V28 Mapping

ICD-10 CODES	HCC (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
L10.0 (Pemphigus vulgaris)	**HCC 387** - Pemphigus, Pemphigoid, and Other Specified Autoimmune Skin Disorders	0.406	**Most common type**; begins in mouth; document mucosal involvement explicitly

AAVBC Knowledge Hub Premium Content Access

Continue reading your guide with an AAVBC subscription

Subscribe Now

Already a subscriber? Sign in

Limited offer

Basic Membership

Free

Includes access to downloadable Quick Reference Guides, mailing list for industry updates/medical information, research toolkits, AAVBC events and so much more!

Cancel at any time