AAVBC

AMERICAN ACADEMY OF VALUE BASED CARE

Pemphigus

Quick Reference Guide

2026

Table of Contents

1. CLINICAL SNAPSHOT **3**
HCC/RAF V28 Mapping **3**
2. RECOGNITION AND DIAGNOSIS **5**
Medicare Screenings and Surveillance (Adult and Geriatric) **5**
Subtle Early Signs in Older Adults (>65) **6**
Geriatric Risk Factors **6**
Diagnostic Thresholds (2020 International Consensus and PDAI) **7**
Clues to Dig Deeper **8**
Common Oversights **8**
Key Differentials in Elderly **9**
Comorbidity Screening **10**
3. MEAT DOCUMENTATION ESSENTIALS **11**
Clinical Documentation Elements **12**
Reframing Common Documentation Shortcuts **13**
4. TREATMENT AND REFERRAL QUICK GUIDE **13**
Therapy Escalation Criteria **14**
Guideline-Aligned Treatment (2020 International Consensus/EDF-EADV) **15**
Non-Pharmacologic Care and Supportive Interventions **16**
Medication Safety and Key Interactions **16**
When to Refer **17**
Follow-Up Timing **18**
Comorbidity Management – Primary Care Role **18**
Cost-Smart Options **19**
Patient Education and Adherence **20**
Quality Metrics Tie-In **21**
5. CODING REMINDERS AND CASE EXAMPLES **23**
Coding Specificity **23**
Annual Clinical Review and Confirmation **24**
Good Documentation - EHR Tips **25**
Brief Case Examples **25**
REFERENCES **26**

1 CLINICAL SNAPSHOT

Definition: Pemphigus is a group of **rare**, potentially life-threatening chronic autoimmune blistering disorders in which **IgG autoantibodies target desmoglein 1 (Dsg1) and/or desmoglein 3 (Dsg3)** - 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-Dsg3 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

ICD-10 CODES	HCC (V28)	RAF (CNA)	DOCUMENTATION REQUIREMENT
L10.1 (Pemphigus vegetans)	**HCC 387**	0.406	**Vegetating variant of PV**; code when documented by specialist
L10.2 (Pemphigus foliaceus)	**HCC 387**	0.406	**Skin only**; no mucosal involvement; document affected body areas
L10.3 (Brazilian pemphigus (fogo selvagem))	**HCC 387**	0.406	**Endemic form**; rarely seen in US Medicare population
L10.4 (Pemphigus erythematosus)	**HCC 387**	0.406	**Overlap with lupus features**; document if specialist-confirmed
L10.5 (Drug-induced pemphigus)	**HCC 387**	0.406	Document **offending drug** or vaccine with adverse effect code; may resolve with discontinuation
L10.81 (Paraneoplastic pemphigus)	**HCC 387**	0.406	**MUST** trigger malignancy workup; code underlying cancer separately
L10.89 (Other pemphigus)	**HCC 387**	0.406	Use only when **subtype documented** but **does not** fit above categories
L12.0 (Pemphigus, unspecified - avoid when subtype is known)	**HCC 387**	0.406	**Do not default to unspecified** if the specialist note identifies the subtype. Query the chart

ABBREVIATIONS: CNA = Community Non-Dual Eligible, Aged (CMS-HCC V28 segment); DIF = direct immunofluorescence; Dsg = desmoglein; HCC = Hierarchical Condition Category; ICD-10 = International Classification of Diseases, 10th Revision; RAF = Risk Adjustment Factor
*RAF values represent the Community Non-Dual Eligible Aged (CNA) coefficient from the 2026 CMS-HCC model; values vary across patient populations based on eligibility and care setting

Risk-Adjusted Care Resources per Patient/Year[8,9]

Risk-adjusted care resource allocation — MA base rate (~$10,402.34) × RAF coefficient

Pemphigus
~$4,223

HCC 387 · RAF 0.406

2 RECOGNITION AND DIAGNOSIS

Medicare Screenings and Surveillance (Adult and Geriatric)

ASSESSMENT	WHEN/FREQUENCY	CLINICAL RATIONALE
Oral mucosa inspection	Any persistent oral erosion **>2-3** weeks unresponsive to empiric therapy[2,11]	**PV begins in the mouth 3 months or longer** before skin lesions in most patients; persistent erosions are the earliest detectable sign[10]
Nikolsky sign	At the bedside whenever pemphigus is suspected[2,10]	Gentle **lateral pressure** shears normal-looking skin in active pemphigus a rapid, office-based clue that warrants biopsy
Dual biopsy (H&E + DIF)[2]	Before starting immunosuppression	**Lesional punch for H&E plus a separate perilesional biopsy in Michel medium for DIF**; DIF is the gold standard with **~77%** sensitivity and **~100%** specificity[12]
Anti-Dsg1/Dsg[3] **ELISA**	At diagnosis and every 3-6 months thereafter	**Detectable in ~98.5%of pemphigus** sera; rising titers predict relapse and guide taper decisions[2,11]
Hepatitis B screen (HBsAg + anti-HBc)[14]	Once before the first rituximab dose[15]	**HBV reactivation** during B-cell depletion can be fatal a mandatory pre-treatment checkpoint[16]
Depression screening (PHQ-9)	At diagnosis and periodically (HEDIS DSF)	Depression affects **28-59%** of PV patients and persists even in clinical remission[17]

ABBREVIATIONS: anti-HBc = hepatitis B core antibody; DIF = direct immunofluorescence; Dsg = desmoglein; ELISA = enzyme-linked immunosorbent assay; H&E = hematoxylin and eosin; HBsAg = hepatitis B surface antigen; HBV = hepatitis B virus; PHQ-9 = Patient Health Questionnaire-9; PV = pemphigus vulgaris

Subtle Early Signs in Older Adults (>65)

EARLY SIGN	WHAT TO LOOK FOR	WHY IT IS MISSED IN OLDER ADULTS
Persistent oral erosions	**Painful, slow-healing erosions** on buccal mucosa, gingiva, or palate; difficulty eating or swallowing[2]	Attributed to **ill-fitting dentures**, **aphthous ulcers, or candidiasis**; antifungal/topical trials delay biopsy[11]
Flaccid skin blisters	Thin-roofed blisters that rupture quickly into raw erosions, often on scalp, trunk, and intertriginous areas[11]	Mistaken for **friction injury, intertrigo, or eczema** in frail or bed-bound patients[2]
Hoarseness or odynophagia	Voice change, throat pain, or sensation of a lump	Laryngeal involvement occurs in **10-40%** of PV; hoarseness is dismissed as reflux or aging voice yet can signal airway risk[18]
Unintentional weight loss	Reduced intake from oral pain; falling serum albumin[2]	Framed as **poor appetite or dementia-related**; albumin <3.5 g/dL independently predicts 10-year mortality **(HR 3.3)**[3]
Conjunctival or genital erosions	Eye irritation, dysuria, or painful mucosal lesions outside the mouth[19]	Patients underreport **genital and ocular symptoms**; clinicians may not examine these sites[2]

ABBREVIATIONS: HR = hazard ratio; PV = pemphigus vulgaris

Geriatric Risk Factors

RISK FACTOR	RISK SIGNAL	CLINICAL RELEVANCE
Age >=65 at diagnosis	**HR 5.9** (5-year) and HR 4.2(10-year) mortality[3]	**Age** is the **strongest independent mortality predictor** across pemphigus cohorts
Hypoalbuminemia (<3.5 g/dL)	**HR 3.3** for 10-year mortality[2,3]	**Reflects severe oral disease limiting intake**; an actionable nutrition target
Diabetes mellitus at diagnosis	**HR 3.8** for 5-year mortality[2,3]	Often steroid-induced; worsens infection risk and wound healing

RISK FACTOR	RISK SIGNAL	CLINICAL RELEVANCE
Neurological disease	**HR 5.4** (5-year) and **HR 5.1** (10-year) mortality[2,3]	Comorbid neurologic illness compounds frailty and care complexity
Lung disease at diagnosis	**HR 3.8** for 10-year mortality[2,3]	**Predisposes to** the **pneumonia** and septicemia that drive inpatient death
Ashkenazi Jewish ancestry	Incidence **~5 per 100,000** vs 0.1-1 per 100,000[2,10]	A recognized epidemiologic association; HLA-DRB1*04:02 enriched in PV

ABBREVIATIONS: HLA = human leukocyte antigen; HR = hazard ratio; PV = pemphigus vulgaris

Diagnostic Thresholds (2020 International Consensus and PDAI)

DIAGNOSTIC ELEMENT	THRESHOLD/CRITERION	NOTES
Three-domain concordance (2020 International Consensus)[20]	Compatible clinical presentation + histopathology + immunological confirmation	Diagnosis **requires agreement** across all **three domains**; no single test is sufficient alone[11]
Direct immunofluorescence (DIF)	**Perilesional biopsy** in Michel medium; intercellular IgG/C3 deposition[2]	**Gold standard**: **~77%** sensitivity and **~100%** specificity. NOT formalin - the transport medium distinction is critical[12]
Anti-Dsg ELISA	**Anti-Dsg**[3] (mucosal/mucocutaneous PV) and/or anti-Dsg[1] (cutaneous PV, PF)	**Positive in ~98.5%** of sera; titers track disease activity[2]
PDAI severity (Pemphigus Disease Area Index)[21,22]	Mild <15 Moderate 15-44 Severe ≥45 (range 0-250)[2]	Recommended scoring instrument; **ICC 0.91** interrater reliability; primary endpoint in Ritux 3 and PEMPHIX[22]
Laryngeal involvement score	≥4 points (hoarseness 2 + dysphagia 4 + PDAI >18) = **>40%** probability	**Identifies airway risk**; AUC 0.78 and NPV **92.1% at the ≥2-point threshold**[18]

ABBREVIATIONS: AUC = area under the curve; DIF = direct immunofluorescence; Dsg = desmoglein; ELISA = enzyme-linked immunosorbent assay; ICC = intraclass correlation coefficient; NPV = negative predictive value; PDAI = Pemphigus Disease Area Index; PF = pemphigus foliaceus; PV = pemphigus vulgaris

Clues to Dig Deeper

PRESENTATION	CLUE TO DIG DEEPER	ACTION
"Stubborn canker sores" for weeks	Oral erosions unresponsive to antifungal or topical anesthetic **>2-3 weeks**[2]	**Test Nikolsky sign**; order dual biopsy (H&E + perilesional DIF); refer to dermatology[2]
Refractory oral lesions despite immunosuppression	HSV DNA is found in **~30%** of treatment-refractory oral lesions and mimics a flare[2]	**Send HSV PCR before escalating immunosuppression**; treat HSV if positive[23]
New blisters in a patient with a known cancer	Paraneoplastic pemphigus **(L10.81)** carries **~60%** 5-year mortality[2]	**Test anti-plakin antibodies**; coordinate urgent oncology evaluation
Voice change with mouth disease	Hoarseness or dysphagia may signal laryngeal involvement **(10-40%of PV)**[18]	**Apply the laryngeal score**; arrange ENT/airway assessment if elevated

ABBREVIATIONS: DIF = direct immunofluorescence; ENT = ear, nose, and throat; H&E = hematoxylin and eosin; HSV = herpes simplex virus; PCR = polymerase chain reaction; PV = pemphigus vulgaris

Common Oversights

COMMON OVERSIGHT	CONSEQUENCE	BETTER PRACTICE
Treating oral erosions as recurrent aphthae or thrush	Average **5-8 month** diagnostic delay; disease spreads before recognition[13]	Maintain a **low threshold** for Nikolsky testing and biopsy when oral erosions persist beyond **2-3 weeks**[2]
Ordering H&E biopsy only, omitting DIF	**Histology alone is often inconclusive**; diagnosis is missed or delayed[2]	**Always** take a s**eparate perilesional** biopsy in **Michel medium for DIF** - the most common diagnostic error[24]
Conflating pemphigus with bullous pemphigoid	Under-treatment of a higher-mortality disease requiring aggressive therapy[2]	**Distinguish flaccid, shearing pemphigus blisters from tense pemphigoid bullae**; confirm with DIF
Missing laryngeal or airway involvement	Unrecognized airway compromise in **10-40%** of PV patients[2]	Ask about hoarseness and dysphagia; apply the laryngeal score and escalate when elevated

COMMON OVERSIGHT	CONSEQUENCE	BETTER PRACTICE
Overlooking HSV superinfection in refractory lesions	Immunosuppression is escalated against an undetected viral mimic **(~30% harbor HSV DNA)**[2]	**Send HSV PCR for refractory oral lesions**
Underrecognizing depression and steroid myopathy	Untreated mood disorder **(28-59%)** and falls from proximal weakness **(47.5%)**[17,25]	**Screen with PHQ-9**; ask about difficulty rising from a chair and assess fall risk

ABBREVIATIONS: DIF = direct immunofluorescence; H&E = hematoxylin and eosin; HSV = herpes simplex virus; PCR = polymerase chain reaction; PHQ-9 = Patient Health Questionnaire-9; PV = pemphigus vulgaris

Key Differentials in Elderly

DIFFERENTIAL	DISTINGUISHING FEATURES	HOW TO CONFIRM
Bullous pemphigoid	**Tense, sturdy subepidermal blisters**; usually spares the mouth; less acantholysis[2]	**DIF shows linear basement-membrane IgG/C3**; anti-BP180/BP230 antibodies[26]
Aphthous stomatitis	Recurrent, self-limited shallow **ulcers without skin blistering** or Nikolsky sign[22,27]	**Clinical course**; **negative DIF and ELISA**
Herpes simplex/herpetic stomatitis	**Grouped vesicles, prodrome**; may also superinfect true pemphigus lesions[27]	**HSV PCR**; recall HSV is found in **~30%** of refractory pemphigus lesions[2]
Erythema multiforme/SJS-TEN	**Target lesions or rapid widespread** detachment with drug exposure[28]	Acute **drug and vaccine history, histology, and absence of intercellular IgG on DIF**
Mucous membrane (cicatricial) pemphigoid	**Scarring conjunctival/oral lesions**; symblepharon[2]	**DIF and target-antigen testing distinguish from pemphigus**

ABBREVIATIONS: DIF = direct immunofluorescence; ELISA = enzyme-linked immunosorbent assay; HSV = herpes simplex virus; PCR = polymerase chain reaction; SJS-TEN = Stevens-Johnson syndrome/toxic epidermal necrolysis

⚠ RED FLAG — EMERGENCY/ACTIVATE CRISIS RESPONSE

- Activate emergency care for any sign of **airway compromise** — hoarseness, stridor, or dysphagia suggesting laryngeal involvement (seen in **10-40%** of PV)
- **Sepsis or hemodynamic instability** from infected erosions — infection is the **leading cause of death**, with over **50%** of hospitalized patients carrying an infection diagnosis
- **Extensive skin denudation** (**>30% body surface area**), which carries burn-unit-equivalent fluid and infection risk
- Do **not** delay transfer to obtain confirmatory testing

Comorbidity Screening

COMORBIDITY	ASSOCIATION IN PEMPHIGUS	SCREENING ACTION
Venous thromboembolism	**~5% first-year VTE risk**; pooled RR **2.17** (95% CI 1.82-2.62); PE adjusted **RR 3.55** in year 1[3,29,30]	Assess **VTE risk at every encounter**, especially during flare, immobilization, or hospitalization
Osteoporosis/fragility fracture	**aOR 2.54** for osteoporosis; **aOR 2.04** for pathological fracture[31]	Baseline **DEXA at steroid initiation**; calcium/vitamin D and bisphosphonate prophylaxis
Steroid-induced diabetes	**21-22% grade 3/4** endocrine events on steroid-only therapy[18]	Monitor **fasting glucose/ HbA1c** at every steroid-dose change; code **E09.65**
Depression and anxiety	Depression **28-59%**; anxiety **25-35.5%**; both persist in remission[17,32]	**PHQ-9 screening**; behavioral-health referral when positive
Steroid-induced myopathy	**47.5% of AIBD patients** on steroids develop proximal weakness[33]	Ask about **difficulty rising from a chair**; PT/OT referral and fall-risk assessment
Serious infection	**>50%** of hospitalized patients carry an infection diagnosis; leading cause of death[7]	**Low threshold for infection workup**; monitor immunoglobulins post-rituximab

COMORBIDITY	ASSOCIATION IN PEMPHIGUS	SCREENING ACTION

ABBREVIATIONS: AIBD = autoimmune blistering disease; aOR = adjusted odds ratio; CI = confidence interval; DEXA = dual-energy X-ray absorptiometry; OT = occupational therapy; PE = pulmonary embolism; PHQ-9 = Patient Health Questionnaire-9; PT = physical therapy; RR = relative risk; VTE = venous thromboembolism

RED FLAG — URGENT (24-72 HOURS)

- Rapidly progressive new blistering
- Suspected **paraneoplastic pemphigus** (**L10.81**, **~60%** 5-year mortality)
- **Ocular involvement** threatening vision
- Signs of **adrenal crisis** during steroid taper (profound fatigue, hypotension, collapse)
- **Oral erosions unresponsive to empiric therapy** beyond **2-3 weeks** — or disease worsening despite adequate immunosuppression (rule out **HSV superinfection**) — warrant urgent dermatology review

AAVBC PERSPECTIVE

*AAVBC encourages primary care providers to recognize that pemphigus vulgaris typically presents in the primary care setting first, as **persistent oral erosions**, often several months before any skin involvement occurs. This diagnostic window is where PCP awareness has the highest clinical yield.*

*AAVBC also supports clear clinical differentiation between pemphigus and **bullous pemphigoid**, which are distinct diseases with different biopsy findings, antibody profiles, mortality risks, and management pathways. Flaccid blisters that rupture on touch, a **positive Nikolsky sign**, and persistent mucosal erosions point toward pemphigus, and tense, sturdy blisters with a negative Nikolsky sign are more consistent with bullous pemphigoid. Conflating these two conditions leads to inappropriate management in a population where both carry meaningful clinical risk. This helps ensure the right patients are identified and referred for the right diagnosis at the right time.*

3 MEAT DOCUMENTATION ESSENTIALS

Pemphigus is a chronic, complex condition whose true severity lives in the secondary complications - malnutrition, steroid-induced diabetes and osteoporosis, infection, and mood disorder. Documentation that captures only "pemphigus" understates the clinical reality. The MEAT framework (Monitor, Evaluate, Assess/Address, Treat) helps the record reflect the active, co-managed nature of the disease at every encounter.

Case vignette: A female patient, 74, on prednisone and maintenance rituximab for biopsy-confirmed pemphigus vulgaris presents for follow-up with new fatigue and two healing oral

erosions. The note records "pemphigus, stable." Over the next quarter she is hospitalized for pneumonia and a vertebral compression fracture. A MEAT-complete note - documenting the anti-Dsg^3 titer trend, albumin, HbA1c at the last steroid change, the DEXA result, immunoglobulin level, and the co-managing dermatologist - would have captured the complexity already present and prompted earlier preventive action.

MONITOR: "Anti-Dsg^3 ELISA: rising trend since last check (repeat q3-6mo). Serum albumin: **3.2 g/dL** (down from 3.9 g/dL last visit) — below the **<3.5 g/dL** mortality-risk threshold. Fasting glucose/HbA1c: not rechecked since last steroid dose increase — overdue. CBC: not indicated (not on azathioprine/mycophenolate). Quantitative IgG: not drawn since last rituximab infusion — overdue. Blood pressure: **138/84** today. New fatigue reported; two healing oral erosions noted on exam."

EVALUATE: "Pemphigus vulgaris, subtype confirmed by prior DIF and anti-Dsg ELISA. Current severity: **PDAI moderate (15-44 range)** based on residual oral erosions and rising Dsg^3 titer trend — rising titers predict relapse and should not be read as reassuring despite clinical improvement. Disease status: **active**, on maintenance immunosuppression (prednisone + rituximab). Falling albumin and unmonitored steroid-related labs suggest clinical complexity beyond what the prior note ('pemphigus, stable')captured."

ASSESS: "Pemphigus vulgaris (**L10.0**), active, on immunosuppression. Steroid-induced diabetes risk: HbA1c overdue since last dose change — order today (**E09.65** if confirmed). Osteoporosis: DEXA not on file — order (**M81.8** + **Z79.52** if confirmed). Malnutrition: albumin **3.2 g/dL** — dietary referral placed (**E44.0** if criteria met). Myopathy: no proximal weakness on exam today (**G72.0** if develops). Depression: **PHQ-9** not administered today — add to next visit (**F32.x** if positive). VTE: no current signs; reassess with any hospitalization. Co-managed with Dr. [dermatology] — last joint visit note reviewed."

TREAT: "Continue prednisone (current dose) and maintenance rituximab per dermatology plan — **Z79.52** (chronic systemic corticosteroids) and **Z79.620** (rituximab) both documented this visit given ongoing treatment burden. Bisphosphonate: not yet started — pending DEXA result. PPI: not currently indicated, reassess with steroid dose. Hepatitis B prophylaxis: status not confirmed — check before next rituximab cycle. Vaccination status: influenza and pneumococcal to be confirmed and updated given immunosuppression. Labs ordered today: HbA1c, quantitative IgG, DEXA referral, PHQ-9 at next visit."

Clinical Documentation Elements

- **Confirmed subtype and method of confirmation** (e.g., "pemphigus vulgaris, DIF-positive, anti-Dsg^3 ELISA-positive")
- **Disease status** (active vs in remission) and current PDAI severity category when available
- **Active immunosuppressive regimen** with long-term medication codes **Z79.52** (steroids) and **Z79.620** (rituximab)
- **Each managed complication with its own code** - diabetes, osteoporosis, malnutrition, myopathy, depression, VTE, infection
- **Co-management with dermatology** and referrals to **nutrition, behavioral health**, and **rehabilitation** services
- **Monitoring results that justify the plan** – titer trend, albumin, HbA1c, DEXA, IgG, and blood pressure

Reframing Common Documentation Shortcuts

DOCUMENTATION SHORTCUT	WHY IT FALLS SHORT	STRONGER FRAMING
"Pemphigus, stable"	**Omits subtype, activity, and the complications driving risk**	"Pemphigus vulgaris (DIF-confirmed), in partial remission on rituximab maintenance; managing steroid-induced diabetes and osteoporosis"
"On steroids"	**Misses the long-term medication code and its toxicity monitoring**	"Long-term systemic corticosteroid use **(Z79.52)**; HbA1c and DEXA monitored; bisphosphonate prophylaxis in place"
"Mouth sores"	**Understates a defining, mortality-relevant disease feature**	"Active oral mucosal erosions limiting intake; albumin **3.2 g/dL**; dietitian and SLP involved"
"Refer to derm"	**Does not reflect ongoing shared management or PCP monitoring role**	"Co-managed with dermatology; PCP monitoring steroid toxicity, infection risk, and bone health between specialist visits"

ABBREVIATIONS: DEXA = dual-energy X-ray absorptiometry; DIF = direct immunofluorescence; Dsg = desmoglein; SLP = speech-language pathology

DOCUMENTATION REMINDER IS COMPREHENSIVE CODING

Documentation that names the **confirmed subtype, the activity status, the active regimen, and each complication** being managed allows an accurate representation of the patient's clinical complexity, and gives the next clinician the context needed to keep the patient safe.

4 TREATMENT AND REFERRAL QUICK GUIDE

Pemphigus care has moved away from prolonged high-dose corticosteroids. For moderate-to-severe disease, current guidelines (**2020 International Consensus** and **EDF/EADV**) recommend **rituximab as first-line therapy** — not a later-stage option.[20,25] In older adult patients, the main

goal is minimizing treatment-related harm while controlling disease, since **treatment complications, not the blisters themselves, are the leading cause of death**. Compared to azathioprine or mycophenolate, rituximab lowers the risk of:

- **Myocardial infarction** (**RR 0.45**)
- **Stroke** (**RR 0.42**)
- **Type 2 diabetes** (**RR 0.63**)
- **Osteoporosis** (**RR 0.46**)

Therapy Escalation Criteria

TRIGGER	ACTION*	RATIONALE
New biopsy-confirmed moderate-to-severe pemphigus	Refer for **first-line rituximab** plus a short **prednisone** course	Ritux 3: **89%** complete remission off steroids at 24 months vs **34%** with prednisone alone[25,30]
Rising anti-Dsg titer or new lesions on maintenance	**Notify dermatology before tapering**; consider rituximab re-dosing	Rising titers predict relapse; PEMPHIX favored rituximab over MMF (**40% vs 10%** sustained remission at 52 weeks)[2,34]
Refractory oral disease despite immunosuppression	**Send HSV PCR before escalating**	HSV DNA is present in **~30%** of refractory oral lesions and mimics a flare[2,23]
Airway, sepsis, or >30% BSA denudation	**Emergency transfer**	Tier 1 emergencies; laryngeal involvement affects **10-40%** of PV[2]

ABBREVIATIONS: BSA = body surface area; Dsg = desmoglein; HSV = herpes simplex virus; MMF = mycophenolate mofetil; PCR = polymerase chain reaction; PV = pemphigus vulgaris ***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Guideline-Aligned Treatment (2020 International Consensus/EDF-EADV)

LINE/SETTING	REGIMEN*	NOTES AND VBC CONSIDERATIONS
First-line (preferred)[2,22,25]	**Rituximab 1000 mg IV** on days 1 and 14, with maintenance 500 mg at months 12 and 18; plus **prednisone 0.5 mg/kg/day** (moderate) or 1.0 mg/kg/day (severe), tapered over 3-6 months	**Halves serious adverse events** and cuts cumulative prednisone by about **two-thirds** vs steroids alone; lower risk of **MI (RR 0.45), stroke (RR 0.42), type 2 diabetes (RR 0.63), and osteoporosis (RR 0.46)** vs azathioprine/MMF[3]
Second-line (rituximab unavailable/ contraindicated)	**Mycophenolate mofetil 2-3 g/day** OR **azathioprine 1-3 mg/kg/day**, adjunctive to corticosteroids[2]	Inferior to rituximab in PEMPHIX. **Check TPMT before azathioprine**; **avoid azathioprine + allopurinol**[34]
Refractory/rescue	**IVIG 2 g/kg over 2-5 days monthly**; cyclophosphamide reserved (high toxicity in elderly) [OFF-LABEL][2]	Expert-consensus level; IVIG preferred over cyclophosphamide in older adults

ABBREVIATIONS: AZA = azathioprine; IV = intravenous; IVIG = intravenous immunoglobulin; MI = myocardial infarction; MMF = mycophenolate mofetil; RR = relative risk; TPMT = thiopurine methyltransferase; VBC = value-based care ***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

CLINICAL PEARL: FIRST-LINE RITUXIMAB PRESERVES FUNCTION

First-line rituximab may seem counterintuitive to clinicians accustomed to steroids-first autoimmune care, but in older adults it is the **function-preserving choice**. In a global cohort of **1,922 patients**, rituximab was associated with substantially lower long-term risk of:

- **Myocardial infarction** (**RR 0.45**)
- **Stroke** (**RR 0.42**)
- **Diabetes** (**RR 0.63**)
- **Osteoporosis** (**RR 0.46**)

(Compared to azathioprine or mycophenolate). The real risk in older adults is the **downstream harm of cumulative steroid exposure**.[3]

Non-Pharmacologic Care and Supportive Interventions

INTERVENTION	INDICATION	GOAL
Specialized wound care	**Open erosions**; fragile, slow-healing skin[2,11]	**Non-adherent/silicone dressings**, gentle handling, moisture-barrier management to prevent secondary infection[35]
Nutrition and SLP support	**Oral erosions limiting intake**; falling albumin[2]	**Soft, nutrient-dense diet**; address dysphagia; prevent malnutrition, since albumin <3.5 g/dL predicts mortality[3]
Bone health	Anticipated steroid use **>3 months**[36]	**Calcium/vitamin D**, weight-bearing activity, and bisphosphonate prophylaxis with baseline DEXA
Physical and occupational therapy	Proximal weakness, fall risk	**Counter steroid-induced myopathy (47.5% of patients)** and preserve independence[33]
Behavioral health and peer support	Depression or anxiety[17]	Counseling/treatment; **the IPPF (ippf.org)** offers peer support and education

ABBREVIATIONS: DEXA = dual-energy X-ray absorptiometry; IPPF = International Pemphigus and Pemphigoid Foundation; SLP = speech-language pathology

Medication Safety and Key Interactions

MEDICATION/PAIR*	CONCERN	ACTION
Rituximab (approved for moderate-to-severe PV)[37]	**HBV reactivation can be fatal**; suppressed vaccine response[38]	**Mandatory HBsAg + anti-HBc screen before first dose**; complete vaccines first; **no live vaccines within 6 months**
Rituximab + live vaccines[38,39]	Contraindicated during B-cell depletion	Use **recombinant zoster** (Shingrix) and **inactivated influenza**; **avoid live vaccines**
Corticosteroids + antidiabetic agents[25]	Steroids raise glucose and blunt control	**Monitor fasting glucose/ HbA1c** at initiation and each dose change

MEDICATION/PAIR*	CONCERN	ACTION
Chronic corticosteroids + NSAIDs/anticoagulants[40]	Markedly increased GI bleeding risk	**Add PPI prophylaxis when co-prescribed**; code peptic ulcer disease if present[41]
Azathioprine + allopurinol[42,43]	Xanthine-oxidase inhibition causes severe myelosuppression	**Avoid the combination**; if unavoidable, reduce azathioprine **~75%** and monitor CBC closely[44]
Bisphosphonates (prophylaxis)[45]	Oral agents contraindicated at low eGFR	**Verify renal function before prescribing**; recommended at steroid initiation for bone protection

ABBREVIATIONS: anti-HBc = hepatitis B core antibody; CBC = complete blood count; eGFR = estimated glomerular filtration rate; GI = gastrointestinal; HBsAg = hepatitis B surface antigen; HBV = hepatitis B virus; NSAID = non-steroidal anti-inflammatory drug; PPI = proton pump inhibitor; PV = pemphigus vulgaris
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

When to Refer

REFER TO	WHEN	PURPOSE
Dermatology (autoimmune blistering)	At first suspicion - **urgently**[20]	Confirm diagnosis, lead immunosuppression; **delayed specialty care correlates with worse outcomes**
Emergency department	Airway compromise, sepsis, or **>30%** BSA denudation[2,18]	**Life-threatening** presentations; do not delay for testing
Oncology	Suspected paraneoplastic pemphigus **(L10.81)**	About **60% 5-year mortality**; treat the underlying malignancy[2]
Endocrinology	Refractory steroid-induced diabetes or adrenal-axis concerns[2]	**Optimize glycemic control**; manage taper-related adrenal insufficiency
Nutrition/SLP, PT/OT, behavioral health	Impaired intake, weakness/falls, or positive depression screen[2]	**Multidisciplinary support to preserve function and quality of life**

ABBREVIATIONS: BSA = body surface area; OT = occupational therapy; PT = physical therapy; SLP = speech-language pathology

Follow-Up Timing

PARAMETER	FREQUENCY	THRESHOLD/ACTION
Anti-Dsg ELISA titers	Every **3-6 months**; more often during flare or taper[18]	**Rising titer**: notify dermatology before taper; consider re-dosing
Fasting glucose/HbA1c	**Monthly** during steroid initiation; **every 3 months** on maintenance[2]	**HbA1c ≥6.5%** or glucose ≥ 126 mg/dL = drug-induced diabetes **(E09.65)**
DEXA bone density	Baseline at steroid start; **annually if >7.5 mg/day prednisone**[36]	T-score **≤-2.5**: initiate bisphosphonate; code **M81.8** + **Z79.52**
CBC with differential	Every **2-4 weeks** starting AZA/MMF; every 3 months when stable[46]	Cytopenias: hold the agent and notify the specialist
Quantitative IgG	Every **6-12 months** post-rituximab; sooner if recurrent infection[47]	Low IgG with recurrent infection: consider IVIG replacement
Serum albumin	Every **1-3 months** with active oral disease[3]	Albumin <3.5 g/dL: code **E44.0**; dietitian/SLP referral

ABBREVIATIONS: AZA = azathioprine; CBC = complete blood count; DEXA = dual-energy X-ray absorptiometry; Dsg = desmoglein; ELISA = enzyme-linked immunosorbent assay; IgG = immunoglobulin G; IVIG = intravenous immunoglobulin; MMF = mycophenolate mofetil; SLP = speech-language pathology

Comorbidity Management – Primary Care Role

COMORBIDITY	PRIMARY CARE ROLE
Steroid-induced diabetes	**Monitor and treat hyperglycemia**; adjust agents at dose changes[18]
Osteoporosis	**Order DEXA**; prescribe calcium/vitamin D w/K2 and bisphosphonate **(aOR 2.54)**[48]
Malnutrition/dehydration	**Track albumin and weight**; engage nutrition and SLP, since albumin <3.5 g/dL predicts mortality[3]
Depression/anxiety	**PHQ-9 screening** (depression 28-59%); initiate treatment or refer[17]
Venous thromboembolism	**Assess risk at each visit** (pooled RR 2.17); prophylaxis during immobility/flare[49]

COMORBIDITY	PRIMARY CARE ROLE
Infection surveillance	**Low threshold for workup**; monitor IgG post-rituximab[2]

ABBREVIATIONS: IgG = immunoglobulin G; PHQ-9 = Patient Health Questionnaire-9; SLP = speech-language pathology

Cost-Smart Options

BRAND (EST. COST)	GENERIC (EST. COST)	EST. SAVINGS	COST-SMART TIP
Brand Rituxan (rituximab) 1g IV per infusion (~$9,256/ infusion; initial course = 2 infusions)[50]	**No FDA-approved biosimilar exists for pemphigus vulgaris in the US**[51,52]	N/A	Rituxan is now preferred first-line over prolonged steroids per the Ritux 3 trial and 2023 consensus; billed under **Medicare Part B** (patient pays **20% coinsurance** after deductible); **PCP must document medical necessity** before infusion; screen **HBsAg and anti-HBc** before initiation — antiviral prophylaxis required if positive; verify **immunoglobulin and B-cell counts** at baseline
Brand Rayos (delayed-release prednisone) (~$450–600/month)[53]	Generic **prednisone** (**~$9–15/month**) — all other brands discontinued[54,55]	~$435–585/ month	Generic prednisone is the correct choice for bridging acute flares; used to bridge to rituximab or steroid-sparing agents — **not as long-term monotherapy**; always coordinate taper plan with dermatology; document **Z79.52** for long-term systemic steroid use; in elderly patients, accelerated osteoporosis, hyperglycemia, and cardiovascular risk require active monitoring
Brand Imuran (azathioprine) (~$60–88/month insurance co-pay; cash price significantly higher[56]	Generic **azathioprine** (**~$13–25/month** with discount card)[56,57]	~$35–63/ month	Generic azathioprine is bioequivalent to Imuran; used as steroid-sparing adjunct to allow taper; monitor **CBC and LFTs** for toxicity; no clinical indication favors brand prescribing

BRAND (EST. COST)	GENERIC (EST. COST)	EST. SAVINGS	COST-SMART TIP
Brand CellCept (mycophenolate mofetil 500mg) (avg retail ~$672/month at standard 1g twice-daily dosing)[58]	Generic **mycophenolate mofetil** 500mg (**~$22–45/month** with discount card)[59,60]	~$627–650/ month	Generic MMF is bioequivalent to CellCept; used as steroid-sparing adjunct; GI tolerability issues are common in elderly patients — if intolerance occurs, consider switching to azathioprine rather than reverting to brand; no clinical indication favors brand prescribing
Brand oral dapsone — all oral brands discontinued	Generic oral **dapsone** 100mg (**~$29–55/ month**)[61]	N/A	Generic oral dapsone is an established low-cost steroid-sparing option; check **G6PD status before initiating** — contraindicated in G6PD deficiency; monitor for dose-dependent **hemolytic anemia**
Brand Bactrim DS (trimethoprim/ sulfamethoxazole) — all brands discontinued	Generic **TMP-SMX** (**~$5–15/month**)[62]	N/A	Generic TMP-SMX is required for **PJP prophylaxis** in patients on combination immunosuppression (rituximab plus steroid-sparing agent); negligible cost relative to the hospitalizations it prevents; prescribe proactively — do not wait for the first opportunistic infection

ABBREVIATIONS: IV = intravenous; PV = pemphigus vulgaris; PJP = *Pneumocystis jirovecii* pneumonia; MMF = mycophenolate mofetil; TMP-SMX = trimethoprim/sulfamethoxazole; CBC = complete blood count; LFTs = liver function tests; HBsAg = hepatitis B surface antigen; anti-HBc = hepatitis B core antibody
***Consult FDA labels for the most up-to-date dosage information, contraindications, and drug-drug interactions**

Patient Education and Adherence

- Teach patients to inspect the **mouth and skin daily** and report new blisters or worsening oral erosions **promptly** rather than waiting for the next visit
- Reinforce gentle skin and wound care, a **soft, nutrient-dense diet** when the mouth is sore, and the importance of **never stopping prednisone abruptly** (risk of adrenal crisis)
- Explain that mood changes are common and treatable — depression is prevalent among patients
- All vaccines should be completed **before rituximab**, with **hepatitis B screening required first**, since reactivation can be fatal
- Trusted patient resources include **NIAMS** and the **International Pemphigus and Pemphigoid Foundation** (ippf.org), which offers peer support and education

DOCUMENTATION REMINDER - PATIENT EDUCATION

Document the education provided : flare recognition, steroid-taper safety, wound care, vaccination status, and mental-health discussion. These conversations are part of comprehensive chronic-disease management and belong in the record.

Quality Metrics Tie-In

MEASURE[63,64]	STANDARD	RELEVANCE TO PEMPHIGUS
Medication Reconciliation Post-Discharge (HEDIS TRC sub-measure 4; MIPS #46)	**Denominator:** patients ≥18 seen in outpatient setting within 30 days of inpatient discharge **Numerator:** discharge medication list reconciled with current medication list documented in outpatient record **Exclusions:** none specified	**Drug-induced thrombocytopenia must be excluded as an etiology at every discharge reconciliation**; any agent with thrombocytopenic potential introduced during admission requires explicit documentation of review and clinical rationale for continuation. Reconciliation note must appear in the outpatient record — verbal confirmation at the visit without documentation does not satisfy the measure
Plan All-Cause Readmissions (CMS Stars PCR; MIPS #479)	**Denominator:** enrollees ≥18 with acute inpatient discharge **Numerator:** unplanned readmission within 30 days (lower is better) **Exclusions:** planned readmissions, hospice, psychiatric	**MIPS #479 is group-level only**; individual PCPs cannot submit it directly. ITP bleeding hospitalizations are a primary readmission driver; ensure 5-7 day post-discharge platelet count follow-up is scheduled before the patient leaves the hospital, and document explicit ED-return criteria (platelet threshold, bleeding signs) in the discharge summary

MEASURE[63,64]	STANDARD	RELEVANCE TO PEMPHIGUS
Comprehensive Diabetes Care: Glycemic Status (HEDIS GSD; MIPS #1)	**Denominator:** patients 18-75 with diabetes (≥2 diagnoses on different service dates in measurement period or prior year, or diagnosis + dispensed diabetes medication) **Numerator (inverse):** most recent HbA1c >9.0% or missing during measurement period **Exclusions:** hospice, palliative care, frailty + advanced illness (≥66)	**MIPS #1 is an inverse measure — a higher rate indicates worse performance.** Corticosteroid-induced hyperglycemia is a direct consequence of high-dose dexamethasone or prednisone induction in ITP. Monitor glucose or HbA1c during and after steroid courses and code any resulting diabetes to specificity (**E08.–** when steroid-induced)
Controlling High Blood Pressure (HEDIS CBP; MIPS #236)	**Denominator:** patients 18-85 with hypertension (≥2 outpatient visits with HTN diagnosis in measurement period or year prior) **Numerator:** most recent BP <140/90 mmHg during measurement period **Exclusions:** ESRD, hospice, palliative care, pregnancy	**Corticosteroids elevate BP through fluid retention and increased vascular resistance**; fostamatinib also causes hypertension as a class effect in a meaningful proportion of treated patients. Monitor BP at every visit during active ITP therapy and document the medication context when BP is elevated to support accurate clinical decision-making

ABBREVIATIONS: BP = blood pressure; CBP = Controlling High Blood Pressure; CMS = Centers for Medicare & Medicaid Services; ESRD = end-stage renal disease; GSD = Glycemic Status Assessment; HbA1c = hemoglobin A1c; HCC = Hierarchical Condition Category; HEDIS = Healthcare Effectiveness Data and Information Set; ITP = immune thrombocytopenia; MIPS = Merit-based Incentive Payment System; PCR = Plan All-Cause Readmissions; TRC = Transitions of Care

AAVBC PERSPECTIVE

AAVBC encourages primary care providers to maintain a low threshold for urgent dermatology referral and to order a **dual biopsy**, a standard H&E biopsy from the blister edge alongside a separate **perilesional direct immunofluorescence (DIF)** biopsy in Michel's medium, at the first sign of persistent mucosal ulcers or fragile blisters that do not respond to standard treatment. DIF is the gold standard for confirming pemphigus; H&E alone is insufficient.
The clinical triggers that should prompt this action include **persistent oral erosions of more than two weeks** not responding to topical treatment, flaccid skin blisters that rupture on touch leaving raw erosions, a positive Nikolsky sign, painful swallowing or throat erosions without a clear infectious cause, and genital or conjunctival erosions occurring alongside oral

or skin findings.
AAVBC supports recognition that **diagnostic delay** in pemphigus allows **progressive mucosal and skin breakdown that increases the risk of sepsis, hospitalization, and potentially fatal outcomes.** Drug-induced pemphigus, triggered by medications including penicillamine and certain antihypertensives, should also be considered and the offending agent discontinued when identified, as resolution may follow without escalating immunosuppression. **Vaccination, including COVID-19 mRNA vaccines,** has also been reported as a trigger or exacerbating factor for pemphigus. When a temporal link is identified, avoidance of revaccination with the same vaccine type should be considered.
Early PCP-level recognition and prompt referral shorten the diagnostic timeline at the point where intervention is most effective. **This helps ensure the right patients receive the right workup and the right referral at the right time.**

QUALITY OUTCOME

In pemphigus, value-based success is measured less by laboratory numbers and more by **patient-reported outcomes**, such as **reduced pain, restored swallowing, better sleep, and psychological well-being**. Proactive primary-care management of steroid toxicity, infection risk, bone health, and mood is the lever that turns accurate documentation into better patient outcomes.

5 CODING REMINDERS AND CASE EXAMPLES

Coding Specificity

CODE	DESCRIPTION	SPECIFICITY/ DOCUMENTATION GUIDANCE
L10.0	Pemphigus vulgaris	**Most common subtype**; confirm with DIF (perilesional, Michel medium) + anti-Dsg3 ELISA; code annually
L10.2	Pemphigus foliaceus	**Superficial**; anti-Dsg1 only; never mucosal - do not document oral involvement for PF
L10.5	Drug-induced pemphigus	**Document the causative agent** and add the appropriate **T36-T50** adverse-effect code

CODE	DESCRIPTION	SPECIFICITY/ DOCUMENTATION GUIDANCE
L10.81	Paraneoplastic pemphigus	**Code the associated malignancy**; anti-plakin antibodies expected
L10.9	Pemphigus, unspecified	**Upgrade to the confirmed subtype once DIF/ELISA return**; unspecified coding underrepresents severity
Z79.52/Z79.620	Long-term systemic steroids/ immunomodulator	**Append at every encounter with ongoing steroid** or rituximab use to capture treatment burden
E44.0/E09.65/M81.8	Malnutrition/drug-induced diabetes/drug-induced osteoporosis	**Frequently omitted secondary complications**; each carries independent risk-adjustment relevance

ABBREVIATIONS: anti-Dsg = anti-desmoglein; DIF = direct immunofluorescence; ELISA = enzyme-linked immunosorbent assay; HCC = Hierarchical Condition Category; PF = pemphigus foliaceus

Annual Clinical Review and Confirmation

- **HCC 387** must be documented and recaptured at a face-to-face encounter every calendar year. Because pemphigus is a chronic condition requiring ongoing immunosuppression, the record should affirm each year that the diagnosis is active or under treatment — not merely carried forward from a problem list
- At each annual review, restate the confirmed subtype and the basis for it (**DIF** and anti-Dsg **ELISA**), the current activity status, and the active regimen with its long-term medication codes. Avoid defaulting to **L10.9** at readmission or follow-up; confirm and recode to the specific subtype
- Use a coding action checklist at every pemphigus encounter: confirm the **L10.x** subtype; append **Z79.52** and/or **Z79.620**; and screen for and code, when present: **E09.65** (drug-induced diabetes), **M81.8** with **Z79.52** (osteoporosis), **E44.0**/**E86.0** (malnutrition/ dehydration), **G72.0** (myopathy), **F32.x**/**F41.x** (mood), VTE (**I82.4x**/**I26.99**), and any active infection (**B00.x**, **B37.x**, **J18.x**, **A41.x**)
- Frame the documentation as an accurate representation of clinical complexity. The secondary codes are not add-ons for scoring — they describe the real disease burden of an older adult on chronic immunosuppression and ensure the next clinician understands what is being managed

Good Documentation - EHR Tips

EHR TIP	HOW	BENEFIT
Subtype-specific problem entry	Enter **L10.0** (or confirmed subtype) rather than **L10.9** once DIF/ELISA return	Preserves specificity and supports accurate annual recapture
Long-term medication smart list	Link **Z79.52/Z79.620** to the active steroid/rituximab orders	Captures treatment burden at every encounter
Monitoring flowsheet	Track titers, albumin, HbA1c, DEXA, IgG, and BP over time	Surfaces the data that justify the plan and the complication codes
Annual documentation reminder	Set a yearly prompt to confirm HCC 387 at a face-to-face visit	Prevents the chronic diagnosis from lapsing

ABBREVIATIONS: BP = blood pressure; DEXA = dual-energy X-ray absorptiometry; DIF = direct immunofluorescence; ELISA = enzyme-linked immunosorbent assay; HCC = Hierarchical Condition Category; IgG = immunoglobulin G

Brief Case Examples

SUCCESS - Early Biopsy, Confirmed Subtype, Steroid-Sparing Care

Patient: Male patient, 68, with 3 weeks of painful oral erosions unresponsive to antifungal therapy.

Documentation: PCP tested Nikolsky sign (positive), ordered dual biopsy (H&E + perilesional DIF in Michel medium), and referred urgently to dermatology. DIF and anti-Dsg3 ELISA confirmed pemphigus vulgaris; note recorded subtype, activity, and co-management.

Outcome: First-line rituximab started; hepatitis B screened first and vaccines completed. Steroid course kept short; HbA1c, DEXA, and albumin monitored. Disease controlled with minimal steroid toxicity.

Coding: L10.0 documented annually with **Z79.52** and **Z79.620**; baseline DEXA documented – an accurate, complete picture of complexity.

PITFALL - Unspecified Coding and Omitted Complexity

Documentation as written: "Mouth sores - thrush. Pemphigus, stable. Continue steroids."

Consequence: Subtype never confirmed; H&E ordered without DIF; complications (steroid-induced diabetes, osteoporosis, falling albumin) managed but never coded. Patient later hospitalized for pneumonia and a vertebral fracture.

Risk-adjustment impact: Carried as **L10.9**; secondary complications omitted - the record understated a high-complexity patient and lost continuity for the next clinician.

Fix: Confirm subtype by DIF/ELISA and recode to **L10.0**; append **Z79.52/Z79.620**; code **E09.65, M81.8**, and **E44.0** as managed; supports HCC 387 annually.

REFERENCES

1. Kasperkiewicz M, Ellebrecht CT, Takahashi H, et al. Pemphigus. *Nat Rev Dis Primers.* 2017;3:17026. doi:10.1038/nrdp.2017.26
2. Schmidt E, Kasperkiewicz M, Joly P. Pemphigus. *Lancet.* 2019;394(10201):882-894. doi:10.1016/S0140-6736(19)31778-7
3. Kridin K, Sagi SZ, Bergman R. Mortality and cause of death in patients with pemphigus. *Acta Derm Venereol.*2017;97(5):607-611.
4. Centers for Medicare & Medicaid Services; National Center for Health Statistics. ICD-10-CM Official Guidelines for Coding and Reporting FY 2026. US Department of Health and Human Services; 2025. Accessed June 9, 2026. https://www.cms.gov/files/document/fy-2026-icd-10-cm-coding-guidelines.pdf
5. Wertenteil S, Garg A, Strunk A, Alloo A. Prevalence estimates for pemphigus in the United States: a sex- and age-adjusted population analysis. *JAMA Dermatol.* 2019;155(5):627-629. doi:10.1001/jamadermatol.2018.5954
6. Langan SM, Smeeth L, Hubbard R, et al. Bullous pemphigoid and pemphigus vulgaris — incidence and mortality in the UK: population based cohort study. *BMJ.* 2008;337:a180.
7. Ren Z, Narla S, Hsu DY, Silverberg JI. Association of serious infections with pemphigus and pemphigoid: analysis of the Nationwide Inpatient Sample. *J Eur Acad Dermatol Venereol.* 2018;32(10):1768-1776.
8. Centers for Medicare & Medicaid Services. 2026 final ICD-10-CM mappings (CMS-HCC V28). CMS; 2025. Accessed July 7, 2026. https://www.cms.gov/medicare/health-plans/medicareadvtgspecratestats/risk-adjustors/2026-model-software
9. Centers for Medicare & Medicaid Services. CMS-HCC model relative factor tables (community, non-dual, aged). CMS; 2024. Accessed July 7, 2026. https://www.cms.gov/files/document/revised-cms-hcc-model-relative-factor-tables.pdf
10. Leuci S, Ruoppo E, Adamo D, Calabria E, Mignogna MD. Oral autoimmune vesicobullous diseases: classification, clinical presentations, molecular mechanisms, diagnostic algorithms, and management. *Periodontol 2000.*2019;80(1):193-225.
11. Bystryn JC, Rudolph JL. Pemphigus. *Lancet.* 2005;366(9479):61-73.
12. Wu N, Cai Y, Wu F, et al. C3d immunohistochemical staining on paraffin-embedded tissue for diagnosis of pemphigus. *Arch Pathol Lab Med.* 2024;148(9):1021-1028.
13. Petruzzi M, Lucchese A, Contaldo M, et al. ELISA detection of anti-desmoglein 1 and anti-desmoglein 3 and indirect immunofluorescence in oral pemphigus: a retrospective study. *Oral Dis.* 2022;28(4):1098-1106.
14. Jeng WJ, Yip TCF, Lok AS. Hepatitis B: a review. *JAMA.* 2026;335(21):1879-1892. doi:10.1001/jama.2026.6070
15. Ruxience (rituximab-pvvr) [prescribing information]. Pfizer Laboratories Div Pfizer Inc; 2024. Accessed July 7, 2026. https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=f941fc61-f7a3-4e4a-ab7c-87c1667fa05b

16. Terrault NA, Lok ASF, McMahon BJ, et al. Update on prevention, diagnosis, and treatment of chronic hepatitis B: AASLD 2018 hepatitis B guidance. *Hepatology.* 2018;67(4):1560-1599. doi:10.1002/hep.29800

17. Pourali SP, Gutierrez Y, Kucharik AH, et al. Bullous dermatoses and depression: a systematic review. *JAMA Dermatol.* 2021;157(12):1487-1496.

18. Ariel D, Tsur N, Mimouni D, et al. Predicting laryngeal involvement in pemphigus vulgaris using a clinical risk score. *JAMA Dermatol.* 2026;162(4):e1-e8.

19. Kurzeja M, Olszewska M, Grzybowski A, Rudnicka L. Ocular involvement in autoimmune bullous diseases. *Clin Dermatol.* 2023;41(5):543-551.

20. Murrell DF, Peña S, Joly P, et al. Diagnosis and management of pemphigus: recommendations of an international panel of experts. *J Am Acad Dermatol.* 2020;82(3):575-585.e1. doi:10.1016/j.jaad.2018.02.021

21. Rosenbach M, Murrell DF, Bystryn JC, et al. Reliability and convergent validity of two outcome instruments for pemphigus. *J Invest Dermatol.* 2009;129(10):2404-2410. doi:10.1038/jid.2009.72

22. Hébert V, Boulard C, Houivet E, et al. Large international validation of ABSIS and PDAI pemphigus severity scores. *J Invest Dermatol.* 2019;139(1):31-37.

23. Baum S, Atar I, Coster D, et al. Relationship between pemphigus vulgaris severity and PCR-positive herpes simplex virus. *Acta Derm Venereol.* 2022;102:adv00710.

24. Reimann JDR, Moynihan SP, Horn TD. Assessment of clinical and laboratory use of the cutaneous direct immunofluorescence assay. *JAMA Dermatol.* 2021;157(11):1340-1345.

25. Joly P, Maho-Vaillant M, Prost-Squarcioni C, et al. First-line rituximab combined with short-term prednisone versus prednisone alone for the treatment of pemphigus (Ritux 3). *Lancet.* 2017;389(10083):2031-2040. doi:10.1016/S0140-6736(17)30070-3

26. Schmidt E, Zillikens D. Pemphigoid diseases. *Lancet.* 2013;381(9863):320-332.

27. Stoopler ET, Villa A, Bindakhil M, Díaz DLO, Sollecito TP. Common oral conditions: a review. *JAMA.* 2024;331(12):1045-1057.

28. Duong TA, Valeyrie-Allanore L, Wolkenstein P, Chosidow O. Severe cutaneous adverse reactions to drugs. *Lancet.*2017;390(10106):1996-2011.

29. Leshem YA, Atzmony L, Dudkiewicz I, Hodak E, Mimouni D. Venous thromboembolism in patients with pemphigus: a cohort study. *J Am Acad Dermatol.* 2017;77(2):340-343.

30. Chen TL, Huang WT, Loh CH, Huang HK, Chi CC. Risk of incident venous thromboembolism among patients with bullous pemphigoid or pemphigus vulgaris: a nationwide cohort study with meta-analysis. *J Am Heart Assoc.*2023;12(17):e030037.

31. Chovatiya R, Silverberg JI. Association of pemphigus and pemphigoid with osteoporosis and pathological fractures. *Arch Dermatol Res.* 2020;312(4):263-271.

32. Wang J, Wu H, Cong W, et al. Psychological morbidity in patients with pemphigus and its clinicodemographic risk factor: a comparative study. *J Dermatol.* 2023;50(10):1271-1280.

33. He A, Koszegi B, Uzun S, et al. Autoimmune blistering diseases treated with glucocorticoids: an international study of steroid-induced myopathy. *J Eur Acad Dermatol Venereol.* 2025;39(2):e1-e8.

34. Werth VP, Joly P, Mimouni D, et al. Rituximab versus mycophenolate mofetil in patients with pemphigus vulgaris. *N Engl J Med.* 2021;384(24):2295-2305. doi:10.1056/NEJMoa2028564

35. Maglie R, Didona D, Collina MC, Antiga E, Paganelli A. Optimizing local treatment in pemphigus and pemphigoid: current evidence and unmet needs. *Int J Dermatol.* 2026;65(7):e1-e12.

36. Humphrey MB, Russell L, Danila MI, et al. 2022 American College of Rheumatology guideline for the prevention and treatment of glucocorticoid-induced osteoporosis. *Arthritis Care Res (Hoboken).* 2023;75(12):2405-2419. doi:10.1002/acr.25240

37. Rituxan (rituximab) [prescribing information]. Genentech Inc; 2025. Accessed July 7, 2026. https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=b172773b-3905-4a1c-ad95-bab4b6126563

38. Hwang JP, Feld JJ, Hammond SP, et al. Hepatitis B virus screening and management for patients with cancer prior to therapy: ASCO provisional clinical opinion update. *J Clin Oncol.* 2020;38(31):3698-3715. doi:10.1200/JCO.20.01757

39. National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology: Prevention and Treatment of Cancer-Related Infections. Version 1.2026. NCCN. Accessed July 7, 2026. https://www.nccn.org/guidelines/guidelines-detail?category=3&id=1412

40. Masclee GM, Valkhoff VE, Coloma PM, et al. Risk of upper gastrointestinal bleeding from different drug combinations. *Gastroenterology.* 2014;147(4):784-792.e9.

41. Almadi MA, Lu Y, Alali AA, Barkun AN. Peptic ulcer disease. *Lancet.* 2024;404(10447):68-81.

42. Azathioprine tablets USP [prescribing information]. Alvogen; 2024. Accessed July 7, 2026. https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=2ffa2fce-895d-469e-8667-b078560aec5f

43. Allopurinol tablets USP [prescribing information]. Aurobindo Pharma Limited; 2024. Accessed July 7, 2026. https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=311ca755-c022-4251-97eb-dd9be86a993f

44. KDIGO clinical practice guideline for the care of kidney transplant recipients. *Am J Transplant.* 2009;9(suppl 3):S1-S155.

45. Eastell R, Rosen CJ, Black DM, et al. Pharmacological management of osteoporosis in postmenopausal women: an Endocrine Society clinical practice guideline. *J Clin Endocrinol Metab.* 2019;104(5):1595-1622. doi:10.1210/jc.2019-00221

46. Cartwright SL, Cartwright MS. Health maintenance for adults with neuromuscular diseases on immunosuppression. *Muscle Nerve.* 2019;59(4):397-405.

47. Otani IM, Lehman HK, Jongco AM, et al. Practical guidance for the diagnosis and management of secondary hypogammaglobulinemia: a Work Group report of the AAAAI Primary Immunodeficiency and Altered Immune Response Committees. *J Allergy Clin Immunol.* 2022;149(5):1525-1560.

48. Hsu DY, Brieva J, Sinha AA, Langan SM, Silverberg JI. Comorbidities and inpatient mortality for pemphigus in the U.S.A. *JAMA Dermatol.* 2020;156(8):891-898.

49. Ungprasert P, Srivali N, Kittanamongkolchai W. Risk of venous thromboembolism in patients with pemphigus: a systematic review and meta-analysis. *J Eur Acad Dermatol Venereol.* 2016;30(5):820-824.

50. Rituxan prices, coupons, copay cards & patient assistance. Drugs.com. Accessed July 7, 2026. https://www.drugs.com/price-guide/rituxan

51. Feaster R. 3 rituximab biosimilars worth considering. GoodRx. Accessed July 7, 2026. https://www.goodrx.com/rituxan/biosimilars

52. Teva launches rituximab biosimilar Truxima in US. GaBI Online. Accessed July 7, 2026. https://www.gabionline.net/biosimilars/news/Teva-launches-rituximab-biosimilar-Truxima-in-US

53. Rayos 2026 prices, coupons & savings tips. GoodRx. Accessed July 7, 2026. https://www.goodrx.com/rayos

54. Prednisone 2026 prices, coupons & savings tips. GoodRx. Accessed July 7, 2026. https://www.goodrx.com/prednisone

55. Prednisone prices, coupons, copay cards & patient assistance. Drugs.com. Accessed July 7, 2026. https://www.drugs.com/price-guide/prednisone

56. Azathioprine 2026 prices, coupons & savings tips. GoodRx. Accessed July 7, 2026. https://www.goodrx.com/azathioprine

57. How to help your patients save money on azathioprine: a provider's guide. Medfinder. Accessed July 7, 2026. https://www.medfinder.com/blog/how-to-help-patients-save-money-on-azathioprine-providers-guide

58. CellCept coupons 2026: up to 80% discount. SingleCare. Accessed July 7, 2026. https://www.singlecare.com/prescription/cellcept

59. Mycophenolate mofetil coupons 2026: up to 80% discount. SingleCare. Accessed July 7, 2026. https://www.singlecare.com/prescription/mycophenolate-mofetil

60. Mycophenolate mofetil prices, coupons, copay cards & patient assistance. Drugs.com. Accessed July 7, 2026. https://www.drugs.com/price-guide/mycophenolate-mofetil

61. Dapsone prices, coupons, copay cards & patient assistance. Drugs.com. Accessed July 7, 2026. https://www.drugs.com/price-guide/dapsone

62. Trimethoprim/sulfamethoxazole 2026 prices, coupons & savings tips. GoodRx. Accessed July 7, 2026. https://www.goodrx.com/trimethoprim-sulfamethoxazole

63. National Committee for Quality Assurance. HEDIS MY 2026, Volume 2: Technical Specifications for Health Plans. National Committee for Quality Assurance; 2025. https://www.ncqa.org/hedis/measures/

64. Centers for Medicare & Medicaid Services. Medicare 2026 Part C & D Star Ratings Technical Notes. CMS; 2025. Accessed July 7, 2026. https://www.cms.gov/medicare/health-drug-plans/part-c-d-performance-data

Medical Disclaimer

The information provided by the American Academy of Value-Based Care (AAVBC) in this document, including but not limited to coding guidance, and related content, is intended for educational and informational purposes only. This content is designed to assist healthcare providers, organizations, and professionals in understanding and applying Value-Based Care principles, practices, and regulatory compliance standards.

AAVBC does not provide legal, clinical, or medical advice. The content presented should not be interpreted or relied upon as specific legal, medical, clinical, or professional guidance. While efforts are made to ensure the accuracy and currency of the information provided, AAVBC does not guarantee that the materials presented are complete, comprehensive, or without error.

Providers and healthcare professionals must independently evaluate all content and guidance presented herein in the context of applicable laws, regulations, clinical guidelines, payer policies, patient-specific conditions, and contraindications. Any treatments, procedures, diagnostic

approaches, medications, or strategies discussed are illustrative and should not be directly applied without a thorough and independent review of current, evidence-based clinical resources, and regulatory requirements.

For coding, documentation, utilization management, quality measures (including STAR ratings), and compliance matters, users are responsible for consulting official guidelines issued by regulatory bodies, including but not limited to the Centers for Medicare and Medicaid Services (CMS), the American Medical Association (AMA), relevant state agencies, and other authoritative entities.

AAVBC expressly disclaims liability for any loss, claim, or damages arising directly or indirectly from the use or reliance on information provided in this document. Users should consult their organization's legal counsel, compliance officer, or qualified professional advisor for advice specific to their situation. By accessing and using this document, you agree to AAVBC's terms and acknowledge that the use of the content is at your own risk and discretion.