

“Pemphigus Vulgaris: An Integrated Review of Clinical Management and Nursing Implications”

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ABSTRACT

Pemphigus vulgaris (PV) is a rare, chronic autoimmune blistering disorder characterized by intraepidermal blister formation and painful erosions of the skin and mucous membranes. The disease can significantly affect nutrition, physical functioning, psychological well-being, and quality of life. Early diagnosis and appropriate treatment are essential to control disease activity and prevent complications. This review aims to provide an integrated overview of pemphigus vulgaris, focusing on its pathophysiology, clinical manifestations, diagnosis, treatment, complications, and implications for nursing practice. Management of PV requires individualized and multidisciplinary care. Pharmacological therapy aims to achieve disease control while minimizing treatment-related adverse effects.

Nurses have a vital role in assessing disease progression, maintaining skin and mucosal integrity, preventing infection, managing pain, supporting nutrition, monitoring treatment-related complications, and promoting medication adherence. Patient and caregiver education is essential for effective self-management and early recognition of complications. Integrated nursing care is essential in improving clinical outcomes, preventing complications, supporting treatment adherence, and enhancing quality of life among individuals with pemphigus vulgaris.

METHODOLOGY OF THE LITERATURE SEARCH

This study was conducted as a narrative and integrated literature review to synthesize current evidence concerning the clinical management and nursing implications of pemphigus vulgaris.

Databases Searched: PubMed, MEDLINE, Scopus, Google Scholar, and the Cumulative Index to Nursing and Allied Health Literature (CINAHL).

Search Period: Literature published between January 2015 and June 2026 was primarily evaluated to capture recent therapeutic paradigms, with foundational immunological texts included as appropriate.

Keywords and Search Strings: "Pemphigus vulgaris", "autoimmune blistering disease", "desmoglein-3", "rituximab therapy in pemphigus", "corticosteroid-sparing immunosuppressants", and "nursing care pathway for pemphigus".

Eligibility Criteria

- Inclusion Criteria: Peer-reviewed consensus guidelines, systematic reviews, randomized controlled trials, observational studies focusing on human subjects, and clinical nursing management articles published in English.

- **Exclusion Criteria:** Animal studies, non-peer-reviewed commentaries, and articles lacking direct clinical relevance to PV diagnosis, medical management, or nursing interventions.

Keywords: Pemphigus vulgaris; autoimmune blistering disease; nursing implications; patient education.

INTRODUCTION

Pemphigus vulgaris (PV) is a rare, chronic, potentially life-threatening autoimmune blistering disorder characterized by the development of fragile intraepidermal blisters and painful erosions involving the mucous membranes and skin. The disease occurs as a consequence of an autoimmune response directed against desmosomal adhesion proteins, particularly desmoglein-3, with desmoglein-1 involvement contributing to cutaneous manifestations. Disruption of keratinocyte adhesion results in acantholysis and subsequent formation of intraepidermal blisters. Although PV is uncommon, it is associated with substantial morbidity because extensive erosions can compromise the protective skin barrier, increase the risk of secondary infection, interfere with oral intake, and adversely affect physical and psychological well-being.^{1,2}

The clinical presentation of PV is variable, but mucosal involvement is particularly characteristic. Oral lesions may be the initial manifestation and commonly present as painful erosions that can interfere with eating, drinking, swallowing, and oral hygiene. Cutaneous disease is characterized by flaccid bullae that rupture easily, leaving extensive erosions and crusted lesions. Because the lesions may initially resemble other ulcerative, infectious, or autoimmune conditions, early recognition and appropriate diagnostic evaluation are essential. Histopathological examination, direct immunofluorescence, and serological assessment of desmoglein autoantibodies are important components of diagnostic confirmation.^{3,4}

The pathogenesis of PV involves a complex interaction between genetic susceptibility, loss of immune tolerance, autoreactive B- and T-cell responses, and production of pathogenic autoantibodies. Anti-desmoglein antibodies interfere with desmosomal adhesion and contribute to keratinocyte separation. Recent evidence has also demonstrated the involvement of broader immune pathways, including T-cell, B-cell, and innate immune responses, suggesting that PV is not solely a disease of antibody-mediated tissue injury but involves a complex immunological network.⁴

Despite advances in treatment, patients with PV frequently require prolonged therapy and continuous monitoring. Complications may arise from both the disease itself and the medications used for its management. Extensive erosions can lead to pain, impaired mobility, fluid and protein loss, secondary infection, and nutritional compromise, whereas prolonged corticosteroid and immunosuppressive therapy may increase susceptibility to infection and cause metabolic, gastrointestinal, cardiovascular, and other adverse effects. Consequently, comprehensive management extends beyond pharmacological treatment and requires coordinated multidisciplinary care.^{1,2}

Nurses have a pivotal role in the holistic management of patients with PV. Nursing care encompasses systematic assessment of skin and mucosal lesions, wound and skin care, pain assessment and management, infection prevention, nutritional support, oral hygiene, medication administration and monitoring, and identification of treatment-related adverse effects.

Nurses also provide essential psychological support and patient education because the chronic and visible nature of PV may contribute to anxiety, social isolation, emotional distress, and reduced quality of life. Effective education regarding medication adherence, wound care, oral care, infection prevention, recognition of warning signs, and follow-up is essential for promoting self-management and preventing complications.⁵

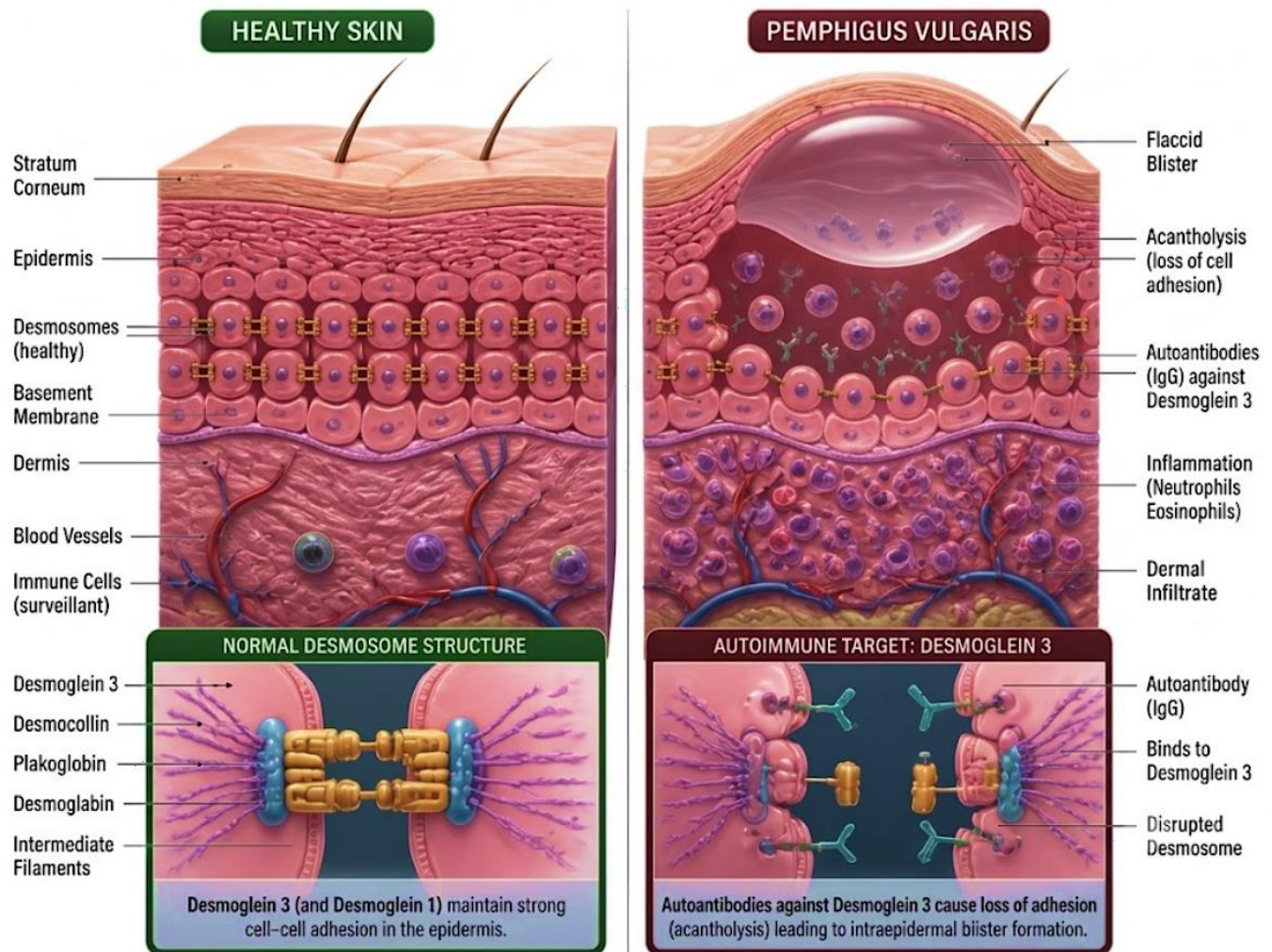


Fig: 1 Overview of Pemphigus Vulgaris

ETIOPATHOGENESIS

Pemphigus vulgaris (PV) is an autoimmune blistering disorder caused by the loss of adhesion between epidermal keratinocytes, a process known as acantholysis. The disease results primarily from pathogenic IgG autoantibodies directed against desmoglein-3 (Dsg3), a cadherin protein located in desmosomes that maintains adhesion between keratinocytes.

Genetic predisposition

Certain HLA class II alleles are associated with increased susceptibility to PV. Genetic predisposition influences abnormal immune recognition of desmosomal proteins, although genetic factors alone are insufficient to cause the disease.¹

Autoimmune response

In susceptible individuals, immune tolerance to desmosomal proteins is disrupted. B lymphocytes produce IgG autoantibodies, predominantly against Dsg3. In patients with both mucosal and cutaneous disease, antibodies against Dsg1 may also develop.²

Role of desmogleins

Desmoglein-3 is highly expressed in the lower layers of the epidermis and mucosal epithelium, whereas desmoglein-1 is more prominent in the superficial epidermis. Autoantibody binding interferes with desmosomal adhesion and intracellular signaling.²

Acantholysis and blister formation

Autoantibodies interfere with keratinocyte adhesion, resulting in acantholysis. Keratinocytes separate from one another, producing an intraepidermal, suprabasal blister. This accounts for the characteristic flaccid blisters and erosions seen clinically.³

Complement and inflammatory mediators

Although direct antibody-mediated disruption of desmosomal function is central, complement activation and inflammatory pathways can contribute to tissue injury. Cytokines and other inflammatory mediators may further promote keratinocyte damage and blister formation.

Environmental and triggering factors

The onset or exacerbation of PV may be associated with factors such as certain medications, infections, physical trauma, ultraviolet exposure, and psychological stress in susceptible individuals. Drug-induced pemphigus is particularly associated with some thiol- or sulfhydryl-containing medications.⁵

Clinical Manifestation

Pemphigus vulgaris (PV) is characterized by mucosal and/or cutaneous blistering and erosions resulting from autoimmune disruption of keratinocyte adhesion. The clinical presentation may vary according to the distribution and profile of desmoglein autoantibodies. Mucosal involvement is common and may precede skin manifestations, while some patients develop predominantly cutaneous disease. The lesions are typically fragile and rupture easily, so patients often present with erosions rather than intact blisters.^{3,4,5}

Positive Nikolsky sign:

A characteristic clinical feature is the Nikolsky sign, in which gentle lateral pressure or rubbing of apparently normal-appearing skin adjacent to a lesion may produce epidermal separation. A positive Nikolsky sign reflects the fragility of epidermal adhesion associated with acantholysis. However, it should be interpreted together with other clinical and diagnostic findings rather than being used alone to establish the diagnosis.²

CLINICAL MANIFESTATION	DESCRIPTION	NURSING IMPLICATIONS
Flaccid blisters	Fragile, easily ruptured blisters containing clear or serous fluid	Handle skin gently and prevent friction or trauma
Skin erosions	Raw, painful areas following rupture of blisters	Assess wound condition, exudate, pain and healing
Mucosal lesions	Commonly involve oral and other mucosal surfaces	Assess oral integrity, swallowing and nutritional intake
Oral pain	Pain associated with erosions and ulceration	Assess pain and provide prescribed analgesia and oral care
Odynophagia	Painful swallowing due to mucosal involvement	Monitor oral intake and hydration; consider dietitian referral
Extensive skin involvement	Widespread erosions may cause significant discomfort and fluid/protein loss	Monitor hydration, nutrition, skin integrity and infection
Positive Nikolsky sign	Epidermis may detach with gentle pressure	Avoid unnecessary pressure and friction during care

Pruritus/burning	May occur around affected areas	Assess severity and provide appropriate symptom management
Secondary infection	Open erosions may facilitate bacterial infection	Monitor for erythema, purulent discharge, fever and systemic symptoms
Psychological distress	Chronic visible lesions and treatment burden may affect emotional well-being	Assess anxiety, depression, social withdrawal and body-image concerns

Table: 1 Clinical Manifestations of Pemphigus Vulgaris and Nursing Implications^{2,3}

Diagnostic Approaches

Clinical Assessment Diagnosis begins with a detailed examination of the skin and mucous membranes. Typical findings include painful oral erosions, fragile flaccid bullae, erosions, and a positive Nikolsky sign. Clinical findings raise suspicion but should be confirmed by laboratory investigations.

Histopathological Examination A biopsy from the edge of a fresh blister is examined using hematoxylin and eosin staining. The characteristic finding in PV is suprabasal intraepidermal separation with acantholysis, often producing the classical “row of tombstones” appearance of basal keratinocytes.

Direct Immunofluorescence (DIF) DIF is a key confirmatory investigation. A biopsy is obtained from clinically normal perilesional skin or mucosa adjacent to a fresh lesion. PV typically demonstrates intercellular IgG and/or C3 deposition in a characteristic “fish-net” or “chicken-wire” pattern.

Indirect Immunofluorescence (IIF) IIF detects circulating autoantibodies in the patient's serum. It can support the diagnosis of pemphigus and may be performed using substrates such as monkey or human esophagus. Although useful, IIF is generally considered complementary when DIF and antigen-specific testing are available.

Desmoglein 1 and 3 ELISA ELISA detects circulating IgG autoantibodies against desmoglein-1 (Dsg1) and desmoglein-3 (Dsg3). Anti-Dsg3 antibodies are strongly associated with mucosal disease, whereas antibodies against both Dsg1 and Dsg3 are commonly associated with mucocutaneous disease. ELISA can also be useful for monitoring antibody levels during treatment.

Tzanck Smear A Tzanck smear is a rapid cytological test performed from the base of a fresh blister or erosion. The presence of acantholytic keratinocytes can support the diagnosis. However, it is not sufficiently specific to replace histopathology and immunofluorescence.

Biochip and Multiparametric Immunoassays Biochip technology can detect several autoantibodies simultaneously, including antibodies against Dsg1 and Dsg3. It can be particularly useful in atypical cases and in differentiating PV from other autoimmune blistering disorders.

Management

Management of pemphigus vulgaris (PV) aims to achieve rapid disease control, induce remission, promote healing of existing lesions, prevent complications, minimize treatment-related toxicity, and maintain long-term remission. Because PV is chronic and potentially severe, treatment should be individualized according to disease severity, extent of mucosal and cutaneous involvement, comorbidities, treatment response, and risk of adverse effects.

Current Indian consensus recommendations emphasize an integrated approach involving systemic therapy, appropriate monitoring, wound and oral care, nutritional support, infection prevention, and patient education.^{8,9}

Systemic corticosteroids

Systemic corticosteroids remain an important component of induction therapy because they rapidly suppress autoimmune inflammation and reduce new blister formation. Prednisolone/prednisone is commonly used, with the dose adjusted according to disease severity. For mild PV, approximately 0.5 mg/kg/day may be used, whereas moderate-to-severe disease may require approximately 1 mg/kg/day, with subsequent gradual tapering after adequate disease control. Prolonged corticosteroid exposure should be minimized because of complications such as hyperglycaemia, hypertension, infection, osteoporosis, weight gain, and adrenal suppression.

Rituximab

Rituximab, an anti-CD20 monoclonal antibody, has become an important treatment option for PV and is used particularly in moderate-to-severe disease. It reduces circulating B cells and consequently decreases production of pathogenic autoantibodies. Current guidelines recommend rituximab in combination with systemic corticosteroids for many patients with moderate-to-severe PV, while it may also be considered in selected patients with mild disease.

Steroid-sparing immunosuppressive therapy

Steroid-sparing agents may be used to reduce cumulative corticosteroid exposure. Common options include:

- Azathioprine
- Mycophenolate mofetil
- Mycophenolate sodium
- Cyclophosphamide in selected situations

Treatment selection should consider disease severity, comorbidities, pregnancy potential, infection risk, affordability, and availability. Regular monitoring of complete blood count and liver and renal function is important with immunosuppressive therapy.

Intravenous immunoglobulin and plasmapheresis

Intravenous immunoglobulin (IVIg) and plasmapheresis may be considered in selected patients with severe, refractory, or difficult-to-control disease. They are generally used as additional or rescue therapies rather than routine first-line treatment.

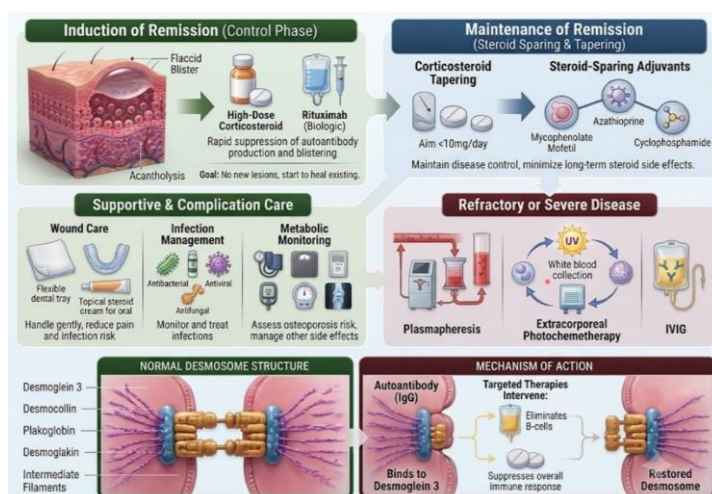


Fig: 2 Comprehensive Management of pemphigus vulgaris

Disease severity	General management approach
Mild/localized PV	Local supportive measures and appropriate systemic therapy based on extent and clinical assessment; topical corticosteroids may be useful for selected lesions
Moderate PV	Systemic corticosteroid-based therapy with consideration of steroid-sparing treatment and/or rituximab
Severe/extensive PV	Systemic corticosteroids with rituximab or other appropriate immunosuppressive therapy; intensive supportive care
Refractory/severe disease	Specialist management; consideration of rituximab, IVIg, plasmapheresis or other advanced therapies according to individual circumstances

Table: 2 Management According to Disease Severity³

Nursing Implications

Nursing management of pemphigus vulgaris (PV) focuses on maintaining skin and mucosal integrity, controlling pain, preventing infection, maintaining adequate nutrition and hydration, monitoring treatment-related adverse effects, providing psychological support, and promoting patient self-management. Because PV is chronic and often requires prolonged immunosuppressive therapy, nursing care should be individualized and coordinated with the multidisciplinary team.^{8,9}

Stage	Nursing action	Desired outcome
1. Initial assessment	Assess skin, mucosa, pain, nutrition, hydration and psychological status	Comprehensive baseline assessment
2. Identify priorities	Determine severity of lesions, pain, infection risk and nutritional compromise	Individualized nursing care plan
3. Skin/wound care	Provide gentle cleansing and appropriate dressing care	Improved skin integrity and comfort
4. Pain management	Assess pain and implement pharmacological/non-pharmacological interventions	Reduced pain
5. Oral care	Assess and maintain oral mucosal health	Improved oral comfort and intake
6. Nutrition/hydration	Monitor intake, weight and hydration	Adequate nutrition and hydration
7. Infection prevention	Maintain appropriate infection-control measures and monitor for infection	Early detection/prevention of infection
8. Medication monitoring	Monitor treatment response, adherence and adverse effects	Safe and effective treatment
9. Psychological support	Assess emotional well-being and provide support/referral	Improved coping
10. Patient education	Teach medication, skin care, oral care, nutrition	Improved self-management

	and warning signs	
11. Discharge planning	Provide individualized home-care instructions and follow-up plan	Safe transition to home
12. Follow-up	Reassess disease activity, complications, adherence and quality of life	Continuity of care and improved outcomes

Table: 3 Nursing Care Pathway for Patients with Pemphigus Vulgaris^{8,9}

Priority Nursing Diagnoses

The following nursing diagnoses may be considered according to the patient's individual assessment:

- Impaired skin integrity related to autoimmune blister formation and epidermal separation.
- Acute pain related to skin and mucosal erosions.
- Impaired oral mucous membrane related to mucosal blistering and erosion.
- Imbalanced nutrition: less than body requirements related to painful oral lesions and difficulty eating.
- Risk for infection related to loss of skin integrity and immunosuppressive therapy.
- Risk for deficient fluid volume related to reduced oral intake and extensive erosions.
- Disturbed sleep pattern related to pain and discomfort.
- Anxiety related to chronic illness, visible lesions, treatment and uncertainty about disease progression.
- Deficient knowledge related to disease process, medication regimen and self-care requirements.
- Ineffective health management related to the complexity and prolonged nature of treatment.

COMPLICATIONS

Disease-Related Complications: Direct consequences of active pathological processes (acantholysis and barrier loss), including extensive fluid and protein loss, electrolyte imbalances, severe dehydration, secondary bacterial cellulitis, sepsis, malnutrition, odynophagia, and site-specific mucosal functional impairments.

Treatment-Related Adverse Effects: Unwanted iatrogenic outcomes driven by therapeutic agents, including corticosteroid-induced hyperglycemia, hypertension, secondary osteoporosis, weight gain, adrenal axis suppression, and immunosuppressive drug-induced cytopenias and opportunistic infections.

Miscellaneous

- **Secondary bacterial infection** – Loss of the protective skin barrier increases susceptibility to bacterial infection and cellulitis.
- **Sepsis** – Severe or untreated secondary infection may progress to systemic infection and sepsis, particularly in extensive disease or immunosuppressed patients.
- **Fluid and electrolyte imbalance** – Extensive erosions can contribute to fluid loss, particularly when a large body surface area is involved.

- **Dehydration** – Painful oral lesions may reduce fluid intake, increasing the risk of dehydration.
- **Malnutrition and weight loss** – Oral pain and difficulty chewing or swallowing can result in inadequate food intake, weight loss, and nutritional deficiencies.
- **Delayed wound healing** – Extensive erosions, infection, nutritional deficiencies, and immunosuppressive treatment can delay tissue repair.
- **Oral complications** – Painful erosions may cause difficulty eating, swallowing, speaking, and maintaining oral hygiene.
- **Mucosal complications** – Involvement of the pharynx, larynx, esophagus, eyes, or genital mucosa may cause site-specific functional problems.
- **Corticosteroid-related complications** – Long-term corticosteroid therapy may cause hyperglycaemia, hypertension, osteoporosis, weight gain, increased infection risk, gastrointestinal complications, and adrenal suppression.
- **Immunosuppressive therapy complications** – Immunosuppressive medications may increase susceptibility to infections and can cause medication-specific hematological, hepatic, renal, or other adverse effects.

CONCLUSION

Pemphigus vulgaris is a chronic autoimmune blistering disorder that significantly affects the skin, mucous membranes, nutrition, and quality of life. Early diagnosis and appropriate treatment are essential to achieve disease control and prevent complications. Comprehensive management includes pharmacological therapy, wound and oral care, pain management, nutritional support, and infection prevention. Nurses play a vital role in assessment, treatment monitoring, patient education, and psychological support. A multidisciplinary, patient-centred approach is essential for promoting healing, preventing complications, and improving quality of life.

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