Autoimmune Skin Conditions

This edition of *FP Essentials*™ will update family physicians on Autoimmune Skin Conditions. The edition will cover the following four topics: psoriasis, pemphigus and pemphigoid, scleroderma, and cutaneous lupus erythematosus.

This edition of *FP Essentials* should be approximately 10,000 words in length, divided into four sections of approximately 2,500 words each (each with an abstract of 200 words or less) plus key practice recommendations, a maximum of 15 tables and figures, suggested reading, and a sufficient number of references (preferably no more than 200) to provide support for all factual statements in the manuscript. This edition should focus on what is new in each topic and should answer the key questions listed for each section. Each section should begin with an illustrative case, similar to the examples provided, with modifications to emphasize key points; each case should have a conclusion that demonstrates resolution of the clinical situation. The references here include information that should be considered in preparation of this edition of *FP Essentials*. However, these references are only a useful starting point that should be used to identify additional information to review.

**Needs Assessment:** Autoimmune skin conditions are a diverse group of conditions caused by immunologic dysregulation that leads to cutaneous and extracutaneous manifestations. They range from common conditions, such as psoriasis, to rare but serious conditions, such as pemphigus and pemphigoid. A survey of members of the American Academy of Family Physicians (AAFP) found significant gaps in medical knowledge and skills related to skin conditions among family physicians, particularly in the evaluation of patients with rashes and systemic conditions with cutaneous manifestations. Some conditions, such as scleroderma, are rated by members as in the top 15% of conditions for which there is a gap between the condition’s relevance to practice and members’ knowledge and skills related to the condition. This edition of *FP Essentials* will discuss the varied clinical presentations and management of psoriasis, the distinguishing features of pemphigus and pemphigoid and guideline-based management, the role of family physicians in the treatment of patients with scleroderma, and evidence-based management of cutaneous lupus erythematosus.
Section 1: Psoriasis

Example case: AB is a 55-year-old man who presents with recurrent pruritic scaly plaques of the scalp, elbows, and knees. Use of dandruff shampoo and thick moisturizing creams has been marginally effective. The lesions are worsening, and now cover significant areas of the extremities and trunk. AB is worried about the costs of therapy, and asks what treatment options are available.

Key questions to consider:

- Briefly discuss the epidemiology and disease burden of psoriasis in the United States, and discuss any racial disparities in severity and disease outcomes.
- What causes psoriasis? Discuss risk factors, provide odds ratios (when available), and succinctly explain its pathophysiology.
- Are there effective primary and secondary preventive interventions for psoriasis?
- How is psoriasis diagnosed? When is biopsy warranted and how should it be performed (eg, punch versus shave, selection of lesions for biopsy)? When are laboratory tests and other tests indicated? When possible, include data on the diagnostic accuracy of suggested clinical and diagnostic tests (eg, clinical findings, biopsy, laboratory tests).
- Which other conditions often are thought to be or are misdiagnosed as psoriasis?
- Using a table (with photographs, if possible), summarize the different types or subtypes of psoriasis and discuss their distinguishing features.
- Provide a treatment algorithm for psoriasis management based on severity (eg, National Psoriasis Foundation classification).
- Among children and adults, when are topical therapies indicated (eg, topical corticosteroids, vitamin D analogue, calcineurin inhibitors, retinoids)? How effective and safe are they?
- Among children and adults, when should systemic therapies be considered (eg, methotrexate, fumaric acid esters, retinoids, biologics)? How effective and safe are they?
- Are there other effective and safe treatments for psoriasis (eg, light therapy, integrative medicine, psychotherapy, behavioral, lifestyle interventions)?
- How should family physicians manage an acute psoriasis flare? How is management different for acute psoriasis than for chronic psoriasis? Why do subspecialists caution against the use of systemic corticosteroids and how strong is the evidence for this?
- What treatment barriers exist for patients with psoriasis who are members of ethnic and racial minority groups? What roles can family physicians and subspecialist physicians play in addressing these barriers?
• Are there any special considerations for psoriasis management in pregnant women?
• When should family physicians consider referring patients to a subspecialist? When is hospitalization warranted?
• Discuss the prognosis of patients with psoriasis, and briefly review nondermatologic complications and their management. Consider presenting this information in a table format.

Initial references to consider:


Section 2: Pemphigus and Pemphigoid

Example case: SH is a 73-year-old woman with uncontrolled type 2 diabetes, a seizure disorder, and heart failure. She presents with a 3-month history of pruritic flaccid blisters, erosions, and crusts. They are located on the central chest, face, scalp, neck, upper back, and bilateral upper and lower extremities. SH has not responded to scabies therapy with two courses of oral corticosteroids. The skin manifestations have returned days after completion of each steroid course. Skin biopsy results reveal intraepidermal acantholysis suggestive of pemphigus foliaceus. SH asks what that means, and what the next steps are for evaluation and treatment.

Key questions to consider:

- What is the difference between pemphigus and pemphigoid?
- Briefly discuss the epidemiology and disease burden of pemphigus and pemphigoid, and the role of family physicians in their evaluation and management.
- Summarize the causes and pathophysiologies of pemphigus and pemphigoid. Discuss risk factors and provide odds ratios (when available).
- Recent literature has suggested an association between bullous pemphigoid and dipeptidyl-peptidase 4 (DPP-4) inhibitors (or gliptins) among patients with diabetes. What should family physicians know about this association? How common is this?
- How are pemphigus and pemphigoid diagnosed? Which clinical findings are helpful and how accurate are they? Consider using figures or tables (with photographs, if available) to summarize the key findings.
- Which other dermatologic conditions should be considered when evaluating patients for suspected pemphigus or pemphigoid (ie, differential diagnosis)?
- What is the role of skin biopsy? How and when should it be performed?
- After confirmation of suggestive histopathologic findings on initial biopsy, discuss the roles of direct immunofluorescence, indirect immunofluorescence, and other serologic tests (eg, immunoassays, enzyme-linked immunosorbent assay [ELISA], immunoblotting) in verifying the diagnoses of pemphigus and pemphigoid. Consider using tables to summarize these concepts, and provide odds ratios (when available) describing the accuracy of diagnostic tests.
- Using figures or tables, review the diagnostic and severity criteria for pemphigus and pemphigoid. Discuss treatment recommendations based on these criteria, and provide strength of evidence information.
- What are the treatment goals for patients with pemphigus and pemphigoid? Briefly discuss why treatment efficacy and safety should be monitored. When can maintenance therapy be discontinued?
- What is the role of topical therapies in management of pemphigus and pemphigoid? How effective and safe are they?
• What are the indications for various systemic therapies for pemphigus and pemphigoid (eg, corticosteroids, immunosuppressants, immunotherapy, immunoglobulins, plasma exchange, dapsone)? How effective and safe are they? Consider using tables to summarize this information.

• What are the complications of pemphigus and pemphigoid? What are the prognoses for patients with these conditions?

Initial references to consider:


Section 3: Scleroderma

Example case: KF is a 50-year-old woman with hypertension, depression, and chronic pain syndrome. She currently smokes half a pack of cigarettes per day. KF presents with a 7-year history of skin thickening of the fingers and hands and fingertip pitting scars. On dermatoscopy, enlarged nail fold capillaries are present. She also reports progressive shortness of breath and dry cough.

Key questions to consider:

- What is scleroderma (ie, systemic sclerosis)?
- Briefly discuss the epidemiology and disease burden of scleroderma in the United States, including any gender or racial disparities in incidence, severity, and outcomes.
- What causes scleroderma? Discuss risk factors, provide odds ratios (when available), and succinctly explain its pathophysiology. Is scleroderma typically diagnosed separately from or along with related conditions?
- How is scleroderma diagnosed? What are the common presenting signs and symptoms? What are the American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) criteria and how accurate is this classification system?
- Discuss the role of the family physician in the evaluation and treatment of patients with scleroderma.
- Which tests are recommended when scleroderma is suspected?
- What are other diagnostic considerations in patients with suspected scleroderma?
- Which other organs (aside from the skin) often are involved in scleroderma? Based on organ involvement, summarize the updated recommendations from the European Alliance of Associations for Rheumatology (EULAR) (formerly the European League Against Rheumatism) for scleroderma management and provide strength of recommendation information.
- What are the treatment goals for patients with scleroderma? Explore the roles of symptomatic treatment, disease control, cure, and palliative care. Is scleroderma curable?
- Discuss the challenges of managing cutaneous fibrosis in scleroderma. Summarize the conventional treatments along with results from promising new trials.
- When is autologous hematopoietic stem cell transplantation indicated? How effective and safe is it?
- Is there a role for surgical, rehabilitative, behavioral, and lifestyle interventions in scleroderma management?
- Given the multiorgan involvement in scleroderma, explain the roles of various subspecialist physicians (eg, dermatology, rheumatology, pulmonology, nephrology subspecialists) in patient care.
• What are the complications and prognoses associated with the various forms of scleroderma? When should hospitalization be considered?

**Initial references to consider:**


Section 4: Cutaneous Lupus Erythematosus

Example case: CG is a healthy 24-year-old man who presents with focal scalp alopecia. It is characterized by a violaceous atrophic plaque, follicular keratotic plugs, adherent scale, and central depigmentation. You obtain a skin sample for biopsy; the results confirm discoid lupus erythematosus.

Key questions to consider:

- What is cutaneous lupus erythematosus (CLE)? How is it related to systemic lupus erythematosus (SLE)?
- How common is CLE? Briefly discuss the epidemiology and disease burden in the United States. Are there gender or racial disparities in the incidence, prevalence, and severity?
- Briefly review the cause of CLE, its pathophysiology, disease course, complications, and associated risk factors.
- What are the common presenting signs and symptoms?
- How is CLE diagnosed? Is there a role for dermatoscopy, and how accurate is it in diagnosing CLE? When is skin biopsy warranted and how should it be obtained? Which serologic tests can aid in the diagnosis?
- What other diagnoses should clinicians consider when evaluating a patient with suspected CLE?
- Discuss treatment goals, including the roles of topical and intralesional therapies, and systemic treatments and their indications.
- Are there roles for surgery and/or physical modalities such as laser and light therapies? If so, when should they be used and how effective are they?
- Are there benefits of lifestyle and behavioral interventions such as smoking cessation and sun protection?
- What is the long-term prognosis for patients with CLE? Do patients ever progress to SLE? If so, how often does this occur? What is the role of biomarkers in predicting progression or other aspects of prognosis?
- When should family physicians consider referring patients with CLE to a dermatology subspecialist?

Initial references to consider:


