Primer on the Rheumatic Diseases

Edited by: John H. Klippel, John H. Stone, Leslie J. Crofford, Patience White

- A tradition of excellence for more than 70 years continues
- Presenting the best translational guide to over 100 rheumatic diseases

revised and EXPANDED

13th EDITION
Primer on the Rheumatic Diseases

Edited by John H. Klippel, Arthritis Foundation, Atlanta, GA, USA
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Leslie J. Crofford, University of Kentucky, Lexington, KY, USA;
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Primer on the Rheumatic Diseases is one of the most prestigious and comprehensive texts on arthritis and related diseases, including osteoarthritis, rheumatoid arthritis, osteoporosis, lupus and more than 100 others. It offers medical students and physicians a concise description of the current science, diagnosis, clinical consequences, and principles of management. New and expanded chapters heighten the translational nature of this edition. Students, trainees, and practicing clinicians all need a standard textbook that can change with the times and reflect recent strides taken in understanding and treating rheumatic disease. The Primer fills that need.

12th ed. published by the Arthritis Foundation, 2001

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New to the 13th Edition:

- New chapters entitled “Clinical Immunology” and “Applied Genetics”, designed to heighten the translational nature of the book.
- A section devoted entirely to juvenile inflammatory arthritis, with individual chapters on “Clinical Features”, “Pathology and Pathogenesis”, “Treatment and Assessment”, and “Special Considerations”.
- Separate chapters on ankylosing spondylitis and the reactive and enteropathic arthropathies, once lumped together (with psoriatic arthritis) as “seronegative spondyloarthropathies”.
- A tripling of the text devoted to psoriatic arthritis, an acknowledgement of the substantial treatment advances in that disorder.
- Individual chapters (and more than doubling of the allotted text) to the metabolic and inflammatory myopathies, once included in the same chapter.
- Reorganization of the vasculitis section along more rational and all-inclusive lines, with a chapter entitled “ANCA-Associated Vasculitis” that addresses together Wegener’s granulomatosis, microscopic polyangiitis, and the Churg-Strauss syndrome, disorders with striking similarities but important contrasts.
- Thoroughly-illustrated chapter related to the cutaneous manifestations of musculoskeletal disease.
- A clinically-focused textbook that addresses the full spectrum of rheumatic disease.
Patients with PG also demonstrate pathergy. This skin thickening has been reported following a variety of surgical procedures or for example, the donor site of a lumbar puncture. The systemic associations vary depending on the type of PG. Classically diagnosed PG and perforating PG are associated more frequently with inflammatory bowel disease and/or arthritis. Cutaneous erosion or inflammatory bowel disease is warranted in cases of peristomal PG, even when the stoma was created for other reasons (e.g., colon cancer surgery). In contrast, atypical pyoderma gangrenosum is found more frequently in the setting of myeloid leukemia or pre-leukemic conditions.

The histopathology of PG is one of ulceration. Although biopsy should be performed to exclude other conditions, PG does have a distinctive histopathology. Because of the importance of including disease mimickers—particularly infections—biopsy is almost always performed as part of the evaluation, despite the possibility that the ulcer will extend through pathergy. Culture of the lesion following skin biopsy is essential. Infectious microorganisms are not excluded but include deep fungal infections, for example, Histoplasmosis, aspergillus, etc. In contrast, histology demonstrates a pyogenic neutrophilic and eosinophilic infiltrate, with papillary edema. Following diagnosis, appropriate antibiotic therapy is employed. Treatment of classical PG includes dapsone (100–200 mg/day), and colchicine. Atypical PG responds less well to dapsone, thalidomide, immunosuppressive agents, and other therapies. Atypical pyoderma gangrenosum is classified as neutrophilic dermatosis of the dorsal hands.

For cases of PG associated with an underlying disease (e.g., inflammatory bowel disease or RA), treatment of the primary condition often leads to improvement in PG. Prednisone (1 mg/kg/day) is generally the first line of therapy for ulcerative PG. Infliximab (3 mg/kg every 8 weeks following two initial dose 2 weeks apart) is also an effective therapy for PG, even in the absence of inflammatory bowel disease. Other therapies employed in PG include dapsone (100–200 mg/day) (assuming normal levels of glucose-6-phosphate dehydrogenase), cyclosporine (5 mg/kg/day), azathioprine (2 mg/kg/day), immunosuppressive agents, and tumor necrosis factor alpha (TNF-alpha) antagonists. Monoclonal antibody–based therapies include dapsone (100–200 mg/day), dapsone, thalidomide, immunosuppressive agents, and other therapies. Atypical PG is classified as neutrophilic dermatosis of the dorsal hands.

### CLINICAL FEATURES

There is substantial overlap in many of the clinical features of the AAVs. In some cases, distinguishing among two or more of these diseases on the basis of clinical features alone is difficult (Table 25E-1).

#### Upper Respiratory Tract and Ears

Although patients with the CSS or MPA may experience substantial ear, nose, or throat disease, this pattern of involvement is most characteristic of WG. More than 90% of patients with WG eventually develop upper airway or ear abnormalities. The nasal symptoms of WG include nasal pain and stuffiness, rhinitis, epistaxis, and secondary crusting. Nasal inflammation may lead to septal erosion, sepal perforation, or, in many cases, nasal bridge collapse—the “Vallecular deformity” (Figure 21C-3). The distinction between active WG in the sinuses and secondary infections in the sinuses may be challenging (see Neomucosal Interventions section).

In 40% to 50% of patients with the CSS, rhinitis is the earliest disease manifestation, typically appearing years prior to the development of full-blown nasal dacryocystitis. Table 21C-3. Clinical features of the primary antineutrophil cytoplasmic antibodies.

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#### TABLE 21C-3. CLINICAL FEATURES OF THE PRIMARY ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED DISORDERS

<table>
<thead>
<tr>
<th>Feature</th>
<th>Wegener's Granulomatosis (WG)</th>
<th>Polyangiitis (PAN)</th>
<th>Microscopic Polyangiitis (MPA)</th>
<th>Neutrophilic Dermatosis</th>
<th>Systemic Lupus Erythematosus (SLE)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Akkasthma</td>
<td>Rare</td>
<td>Common</td>
<td>Rare</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Akkasthma strictly</td>
<td>PR3 / MPO</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arthritis  Krislin</td>
<td>Rare</td>
<td>Common</td>
<td>Common</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Baseline inflammatory vascularity</td>
<td>Common</td>
<td>Common</td>
<td>Common</td>
<td>Common</td>
<td>Rare</td>
</tr>
<tr>
<td>Clinical manifestations</td>
<td>Common</td>
<td>Common</td>
<td>Common</td>
<td>Common</td>
<td>Common</td>
</tr>
<tr>
<td>Eosinophilia</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Erythromelalgia</td>
<td>Occasionally</td>
<td>Occasionally</td>
<td>Occasionally</td>
<td>Common</td>
<td>Common</td>
</tr>
<tr>
<td>Fingers</td>
<td>Saddle-bone deformity</td>
<td>Common</td>
<td>Common</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Gastrointestinal manifestations</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Heart</td>
<td>Occasional valvar dehiscence</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
</tr>
</tbody>
</table>

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**TABLE 25E-1. CLINICAL FEATURES OF THE PRIMARY ANTINEUTROPHIL CYTOPLASMIC ANTIBODIES**

**CLINICAL FEATURES**

There is substantial overlap in many of the clinical features of the AAVs. In some cases, distinguishing among two or more of these diseases on the basis of clinical features alone is difficult (Table 21C-3).
C. Treatment and Assessment
Maya H. Buch and James R. Seibold

B. Epidemiology, Pathology, and Pathogenesis
John Varga; Systemic Sclerosis
- A. Clinical Features
Maureen D. Mayes; Antiphospholipid Syndrome
Michelle Petri

Manzi and Amy H. Kao

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David S. Pisetsky; Patience H. White and Rowland W. Chang

Public Health and Arthritis: A Growing Imperative
Jill P. Buyon; Systemic Lupus Erythematosus
Preeti Jaggi

Steven R. Ytterberg;
D. Mycobacterial, Fungal, and Parasitic Arthritis
Bockensted; Linda K.

C. Lyme Disease
Leonard H. Calabrese;
miscellaneous Crystals
Robert A. Terkeltaub

generative, Pathology, and Pathogenesis
C. Treatment
Hyon K. Choi;
Gout - A. Clinical Features
Dafna D. Gladman;
B. Pathology and Pathogenesis Christopher Ritchlin; C. Treatment and Assessment Philip J. Mease

A. Ankylosing Spondylitis - A. Clinical Features Désirée Van der Heijdt; B. Pathology and Pathogenesis Juergen Braun; C. Treatment and Assessment John C. Davis, Jr.

D. Reactive and Enteropathic Arthritis - Robert D. Inman

A. Osteoarthritis - A. Clinical Features Paul Dieppe;
B. Pathology and Pathogenesis Francis Berenbaum;
C. Treatment Leena Sharma

Gout - A. Clinical Features N. Lawrence Edwards; B. Epidemiology, Pathology, and Pathogenesis Hyon K. Choi; C. Treatment Robert A. Terkeltaub

Calcium Pyrophosphate Dihydrate, Hydroxyapatite, and Miscellaneous Crystals Geraldine McCarthy


Systemic Lupus Erythematosus - A. Clinical and Laboratory Features Jill P. Buyon; B. Epidemiology, Pathology and Pathogenesis David S. Pisetsky; C. Treatment and Assessment Susan Manzi and Amy H. Kao

Antiphospholipid Syndrome - Michelle Petri

Systemic Sclerosis - A. Clinical Features Maureen D. Mayes; B. Epidemiology, Pathology, and Pathogenesis John Varga; C. Treatment and Assessment Maya H. Buch and James R. Seibold

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From the Foreword

The 13th edition of the Primer on the Rheumatic Diseases is an extraordinary handbook for clinical care. The Primer will educate trainees, update established clinicians, and help health care providers from all walks of the profession provide better care for patients with arthritis and rheumatic diseases. I congratulate the editors on their superb work. In addition, the multiple contributors — many of whom are members of the American College of Rheumatology — should be thanked for their scholarly contributions to the Primer. ► Michael E. Weinblatt, MD, Professor of Medicine, Harvard Medical School, Brigham and Women’s Hospital, Boston, MA, USA

About the Editors

John H. Klippel, M.D. is the President and Chief Executive Officer of the Arthritis Foundation. He previously served as a Senior Investigator in the Arthritis and Rheumatism Branch (NIH) (1976-1987), Clinical Director of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) (1987-1999), and Medical Director of the Arthritis Foundation (1999-2003). He is a diplomate of the American Board of Internal Medicine and a fellow of the American College of Physicians and the American College of Rheumatology. His honors and awards include the Surgeon General’s Exemplary Service Award, Distinguished Clinical Teacher Award (NIH Clinical Center), Directors Award (NIH Clinical Center) and the Burroughs-Wellcome Visiting Professor Award from the Royal Society of Medicine in London.

He received a bachelor’s degree from Bowling Green State University and a doctor of medicine degree from the University of Cincinnati College of Medicine. He completed his residency in internal medicine at Yale-New Haven Hospital and his fellowship in rheumatology at the National Institutes of Health and the University of California at San Diego.

John H. Stone, M.D., M.P.H., co-founded and directed the Vasculitis Center at Johns Hopkins University. Dr. Stone attended Harvard Medical School before training in internal medicine at Johns Hopkins and performing his rheumatology fellowship at the University of California-San Francisco. While on the faculty at Johns Hopkins, Dr. Stone served as the Principal Investigator for first randomized clinical trial in Wegener’s granulomatosis in the U.S. and organized the Rituximab in ANCA-Associated Vasculitis trial. From 2002 to 2006, Dr. Stone served as the Deputy Director for Clinical Research at the Johns Hopkins Bayview Medical Center. He was named a Hugh and Renna Cosner Scholar in the Cosner Program on Translational Research (2005). Dr. Stone became Deputy Editor for Rheumatology at UpToDate in 2006 and is an Associate Physician at the Massachusetts General Hospital.

Leslie J. Crofford, M.D. is an active member of the American College of Rheumatology, serving previously as a member of the Committee on Research and Chair of the Committee on Journal Publications. She is currently Vice-President of the American College of Rheumatology Research and Education Foundation and sits on the Executive Committee of the College. Dr. Crofford was elected to the American Board of Internal Medicine for Rheumatology in 2002 and is currently serving her second term. She is on the Board of Trustees of the Ohio River Valley Chapter of the Arthritis Foundation and has served on the Medical and Scientific Committee of the National Arthritis Foundation. Dr. Crofford is active as a clinical rheumatologist and has been named as one of America’s Top Doctors.

Patience White, M.D. is the chief public health officer of the Arthritis Foundation. In addition to her work there, she is a professor of medicine and pediatrics at the George Washington University School of Medicine and Health Sciences and teaches a Health Policy seminar for Stanford University at the Stanford in Washington campus in Washington DC.
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