Primer on the Rheumatic Diseases

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

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revised and EXPANDED
Primer on the Rheumatic Diseases

Edited by John H. Klippel, Arthritis Foundation, Atlanta, GA, USA
Coeditors: John H. Stone, Massachusetts General Hospital, Boston, MA, USA;
Leslie J. Crofford, University of Kentucky, Lexington, KY, USA;
Patience White, Arthritis Foundation, Washington, DC, USA

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.

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13th ed. 2007. 724 p. 124 illus., 111 in color. Softcover
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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 spondyloarthopathies”.
► 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.

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**FIGURE 25E-3**

Patients with PG also demonstrate pathergy. This thin condition has been reported following a variety of surgical procedures, for example, thoracotomy or for clinical reasons. The systemic associations vary depending on the type of PG. Classic disease and perforated PG are associated more frequently with inflammatory bowel disease (IBD) and arthritis. Cutaneous evaluation (or inflammatory bowel disease is warranted in cases of peristomal PG, even when the stoma was created for other reasons (e.g., cancer therapy). In contrast, atypical pyoderma gangrenosum is found more frequently insetting of myeloid leukemias or preleukemic conditions.

Atypical pyoderma gangrenosum is one of the most common complications of PG in use of esacpe. Although biopsies 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 pathology. Cultures of the lesions following skin biopsy is essential. If there is evidence of tuberculosis, leprosy, or other infections, further investigation should be undertaken to exclude inflammatory bowel disease, hamartomatous arthropathy, or sarcoidosis.

**TABLE 25E-1.**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Wegener’s Granulomatosis</th>
<th>Neutrophilic Dermatoses Syndrome of the Dorsal Hands</th>
<th>Sweet’s Syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset</td>
<td>Median</td>
<td>Median</td>
<td>Median</td>
</tr>
<tr>
<td>Gender</td>
<td>Male</td>
<td>Male</td>
<td>Male</td>
</tr>
<tr>
<td>Necrosis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Large vessel vasculitis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Pulmonary hemorrhage</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Microscopic polyangiitis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Alveolar hemorrhage</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Asthma; fleeting infiltrates; hemoptysis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Uveitis</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Ocular ischemia</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Periocular pain</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Neurologic symptoms</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Renal involvement</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
</tbody>
</table>

**FIGURE 25E-4**

Atypical pyoderma gangrenosum, also known as neutrophilic dermatosis of the dorsal hands.

**FIGURE 25E-5**

Monocytic colony-stimulating factor (M-CSF), macrophage colony-stimulating factor (MCP), and tumor necrosis factor alpha (TNF-alpha) antagonists are often used. In patients with acute, idiopathic disease, the prognosis is generally good; many have only one episode. However, the course of patients with underlying leukemias or myelodysplasia follows that of the associated disease. Absent disease remission or a cure, recurrences are common.

**CLINICAL FEATURES**

There is extensive 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-1).

**Upper Respiratory Tract and Ears**

Although patients with the CSS or MPA may experience substantial cutaneous or oral disease, often the most characteristic of WEG. More than 90% of patients with WEG eventually develop upper airway or ear abnormalities. The nasal symptoms of WEG include nasal pain and fullness, rhinorrhea, epistaxis, or nosebleeds. Nasal inflammation may lead to septal erosion, perforation, or, in some cases, nasal bridge collapse—the “vulcanized deformity” (Figure 21C-1). The distinction between active WEG in the sinuses and secondary infections in the maxillo-mandible is challenging (see Nonmedical Interventions section).

In 60% to 75% of patients with the CSS, rhinitis is the earliest disease manifestation, typically appearing years before the development of full-blown sinus disease. Erosive maxillary sinusitis (Figure 21C-1) is generally the first line of therapy for ulcers of WEG, although (5- to 10-day tapering doses every 6 weeks following two initial doses 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 (500 to 1000 mg/day, assuming normal levels of thiopurine methyltransferase [TPMT]), cyclosporine (5 mg/kg/day), azathioprine (2 mg/kg/day, b.i.d.), and sulfasalazine (1.5 g/day). 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 ulcers of WEG, although (5- to 10-day tapering doses every 6 weeks following two initial doses 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 (500 to 1000 mg/day, assuming normal levels of thiopurine methyltransferase [TPMT]), cyclosporine (5 mg/kg/day), azathioprine (2 mg/kg/day, b.i.d.), and sulfasalazine (1.5 g/day).
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 diplomat 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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