Primer on the Rheumatic Diseases

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

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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.

12th ed. published by the Arthritis Foundation, 2001

13th ed. 2007. 724 p. 124 illus., 111 in color. Softcover
ISBN 978-0-387-35664-8 ► $79.95

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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Patients with PG also demonstrate pathergy. This thin condition has been reported following a variety of surgical procedures, for example, thoracotomy or lumbar puncture. The systemic associations vary depending on the type of PG. Classical disease and periostomal PG are associated more frequently with inflammatory bowel disease and/or arthritis. Cutaneous involvement or inflammatory bowel disease is warranted in cases of peristomal PG, even when the stoma was created for other reasons (e.g., following cancer surgery). In contrast, atypical pyoderma gangrenosum is found more frequently in the setting of myeloproliferative or pre-leukemic conditions.

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. Cultures of the lesions following skin biopsy is essential. Infectious lesions are not common but include deep fungal infection, for example, Mucormycosis, aspergillus, histoplasmosis, and coccidioidomycosis, as well as tuberculosis, atypical mycobacteria, and others. Following diagnosis, appropriate antifungal agents are prescribed. In patients with neoplasms, appropriate diagnostics are performed. The clinical associations of PG include granulomatous inflammation (rarely seen in renal biopsy specimens) and ulcerating fibrosclerosis (up to 10% of patients).

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, followed by azathioprine (1–2 mg/kg/day) every 4 weeks following two initial doses (2 weeks each) in a dose-escalating fashion 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 sulfa drugs), methotrexate (7.5 mg/kg weekly), trimethoprim-sulfamethoxazole (150 mg/kg every 24 hours initially, followed by 75 mg/kg every 24 hours), antimalarials (chloroquine, hydroxychloroquine), and hydroxyurea (500 mg daily).

Pyoderma gangrenosum (PG) is a form of ulcerative skin disease. There are at least four clinical variants of PG: classical, atypical, peristomal, and mucosal (3). The classical lesion is a rapidly progressing, painful ulcer, often on the leg, with a violaceous, undermined (overhanging) border (Figure 25E-1). Atypical PG occurs as a more superficial lesion, often on the dorsal hands (Figure 25E-2), extensor forearms, or face. The borders of atypical PG may appear bullous, leading to clinical confusion with Sweet’s syndrome. Peristomal PG occurs as a deep ulcer near the site of a stoma, usually created after gastrointestinal or genitourinary surgery. Finally, mucosal PG is associated with ulcerations that can resemble simple aphthous or vegetative lesions. Mucosal PG must be differentiated from Behcet’s disease.

### Clinical Features

There is considerable 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 upper respiratory limitation, the most characteristic of WG is the presence of a high percentage of patients with WG eventually develop upper respiratory or ear abnormalities. The nasal symptoms of WG include nasal pain and stuffiness, rhinorrhea, rhinorrhea, and oronasal hematoma. Nasal inflammation may lead to septal erosion, perforation, or, in many cases, nasal bridge collapse—the “vallecular deformity” (Figure 21C-3). The distinction between active WG in the sinonasal and secondary infections in the sinonasal lumen may be challenging (see Neutrophilic Inflammation Sections).

In 35% to 55% of patients with the CSS, rhinitis is the earliest disease manifestation, typically appearing years before the development of full-blown disease. LESIONS WITH NEUTROPHILIC DERMATITIS

<table>
<thead>
<tr>
<th>Schwart’s Syndrome</th>
<th>Pyoderma Gangrenosum (PG)</th>
<th>Necrotizing Neutrophilic Dermatosis (NNDH)</th>
<th>Neutrophilic Dermatosis of the Dorsal Hands</th>
</tr>
</thead>
<tbody>
<tr>
<td>Some</td>
<td>Occasional</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>20%-25%</td>
<td>10% for superficial forms</td>
<td>Yes, occasionally</td>
<td>Occasional</td>
</tr>
<tr>
<td>Rare</td>
<td>15% for the superficial forms</td>
<td>No, but joint disease may simulate RA</td>
<td>Occasional</td>
</tr>
<tr>
<td>Bare</td>
<td>Yes, occasionally</td>
<td>No</td>
<td>Occasional</td>
</tr>
<tr>
<td>Bare</td>
<td>15%</td>
<td>No</td>
<td>Occasional</td>
</tr>
<tr>
<td>occasionally</td>
<td></td>
<td>No</td>
<td>Occasional</td>
</tr>
<tr>
<td>Occasionally</td>
<td></td>
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<td>Occasionally</td>
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<td>No</td>
<td>Occasional</td>
</tr>
<tr>
<td>Occasionally</td>
<td></td>
<td>No</td>
<td>Occasional</td>
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</table>

### Table 21C-1

<table>
<thead>
<tr>
<th>Feature</th>
<th>Wegener’s Granulomatosis</th>
<th>Polyarteritis nodosa</th>
<th>Moya-Moya Syndrome</th>
<th>Polyarteritis nodosa</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Eye</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Ear</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Parotid swelling</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Sudden hearing loss</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Mucosal PG</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Cutaneous PG (peristomal)</td>
<td>Occasional</td>
<td>Occasional</td>
<td>Occasional</td>
<td>Occasional</td>
</tr>
<tr>
<td>Nasal polyposis</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Nasal polyps</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Rhinorrhea</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Sinusitis</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
<tr>
<td>Nasal polyposis</td>
<td>Absent or mild</td>
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<tr>
<td>Sudden hearing loss</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
<td>Absent or mild</td>
</tr>
</tbody>
</table>


**ABBREVIATIONS:** ANCA, antineutrophil cytoplasmic antibody; MPO, myeloperoxidase; PR3, proteinase 3.
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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 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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Klippel, J.H.; Stone, J.H.; Crofford, L.E.J.; White, P.H.
(Eds.)
2008, XIX, 721 p., Softcover