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
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.

Available in November 2007

Sample chapters available online at springer.com!

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. Thus, this condition has been reported following a variety of surgical procedures, for example, thoracotomy or fasciotomy. Complete surgical excision of PG is curative. Although biopsy should be performed to exclude other conditions, PG does have a distinctive histopathology. Because of the importance of excluding disease mimics—particularly infections—biopsy is almost always performed as part of the excision, despite the possibility that the ulcer will extend through pathology. Culture of the lesion following skin biopsy is essential. Infections are the most common but include deep fungal infections, for example, Histoplasmosis, coccidioidomycosis, as well as tuberculosis, leishmaniasis, and other fungal diseases. As with Sweet’s syndrome, neutrophilic infiltration of organs other than the skin may also occur in PG. 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.

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, painless ulcer, most often on the leg, with a violaceous, undermined (overhanging) border (Figure 25E-3). Atypical PG occurs as a more superficial lesion, often on the oral or anal margins (Figure 25E-4), external fornices, or face. The borders of atypical PG may appear bluish, 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 gynecologic surgery. Finally, mucosal PG is associated with ulcerations that can resemble simple aphthae or vegetative lesions. Mucosal PG must be differentiated from Behçet’s disease.

**TABLE 25E-1. ASSOCIATIONS WITH NEUTROPHILIC DERMATITIDES.**

<table>
<thead>
<tr>
<th>SWEBT’S SYNDROME</th>
<th>PYODERMA CANCERISGENESIS</th>
<th>BOWEL-ASSOCIATED IMMUNE-MEDIATED DERMATITIS</th>
<th>NEUTROPHILIC DERMATITIS OF THE DIGITAL HANDS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Some</td>
<td>20%-25%</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Often</td>
<td>Yes, occasionally</td>
<td>No</td>
<td>May simulate RA</td>
</tr>
<tr>
<td>Rare</td>
<td>No, joint disease</td>
<td>Rare</td>
<td>No</td>
</tr>
<tr>
<td>Occasionally</td>
<td>No, no possibility</td>
<td>Rare</td>
<td>Occasional</td>
</tr>
</tbody>
</table>

**COLOR FIGURES DEPICT CUTANEOUS FINDINGS AND HISTOPATHOLOGY**

- Figure 25E-1: Sweet’s syndrome.
- Figure 25E-2: Histopathological findings in Sweet’s syndrome.
- Figure 25E-3: Atypical PG.
- Figure 25E-4: Pyoderma gangrenosum.

**CLINICAL FEATURES**

There is relatively little 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 ear, nose, or sinus 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, rhinorrhea, rhinorrhea, and blood or bloody crusts. Nasal inflammation may lead to septal erosions, perforation, or, in many cases, nasal bridge collapse—the "valveless deformity" (Figure 21C-3). The distinction between active WG in the sinus and secondary infections in the sinuses may be 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 signs and symptoms.
### Table of Contents

- **Public Health and Arthritis: A Growing Imperative** - Patience H. White and Rowland W. Chang
- **Evaluation of the Patient - A History and Physical Examination** - David B. Robinson and Hani S. El-Gabalawy; B. Laboratory Assessment - Kerstin Morehead; C. Arthrocentesis, Synovial Fluid Analysis, and Synovial Biopsy - Kenneth H. Fye; D. Imaging of Rheumatologic Diseases - William W. Scott, Jr., William J. Didie, and Laura M. Fayad
- **Signs and Symptoms of Musculoskeletal Disorders** - A. Monarticular Joint Disease - R. Schumacher and Lan X. Chen; B. Polyarticular Joint Disease - Sterling West; C. Neck and Back Pain - D. Regional Rheumatic Pain Syndromes - Joseph J. Biundo, Jr.; E. The Fibromyalgia Syndrome - Dina Dadabhoy and Daniel J. Clauw
- **Molecular and Cellular Basis of Immunity and Immunological Diseases** - Kevin Elias, Richard Siegel, and John J. O'Shea
- **Genetics and Disease** - James Kelley and Robert P. Kimberly
- **Rheumatoid Arthritis** - A. Clinical and Laboratory Manifestations - Christopher V. Tehilian and Joan M. Bathon; B. Epidemiology, Pathology, and Pathogenesis - Jean-Marc Waldenburger and Gary S. Firestein; C. Treatment and Assessment - Alyce M. Oliver and E. William St. Clair
- **Juvenile Idiopathic Arthritis** - A. Clinical Features - Daniel J. Lovell; B. Pathology and Pathogenesis - Patricia Woo; C. Treatment and Assessment - Philip J. Hashkes and Ronald M. Laxer; D. Special Considerations - Carol B. Lindley
- **Psoriatic Arthritis** - A. Clinical Features - Dafna D. Gladman; B. Pathology and Pathogenesis - Christopher Ritchlin; C. Treatment and Assessment - Philip J. Mease
- **Ankylosing Spondylitis** - A. Clinical Features - Désirée Van der Heijdt; B. Pathology and Pathogenesis - Juergen Braun; C. Treatment and Assessment - John C. Davis, Jr.
- **Reactive and Enteropathic Arthritis** - Robert D. Inman
- **Osteoarthritis** - A. Clinical Features - Paul Dieppe; B. Pathology and Pathogenesis - Francis Berenbaum; C. Treatment - Leena Sharma
- **Gout** - A. Clinical Features - 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
- **Idiopathic Inflammatory Myopathies** - A. Clinical Features - Robert L. Wortmann; B. Pathology and Pathogenesis - Lisa G. Rider and Frederick W. Miller; C. Treatment and Assessment - Chester V. Oddis
- **Metabolic Myopathies** - Alan N. Baer
- **Sjögren’s Syndrome** - Troy Daniels
- **Vasculitides** - A. Large Vessel Vasculitis and Polymyalgia Rheumatica - Cornelia M. Weyand and Jörg J. Goronzy; B. Polyal- teritis Nodosa - Keith T. Rott; C. The Antineutrophil Cytoplasmic Antibody–Associated Vasculitides - Wegener’s Granulomatosis, Microscopic Polyangiitis, and the Churg-Strauss Syndrome - John H. Stone; D. Immune Complex-Mediated Vasculitis - Philip Seo; E. Miscellaneous Vasculitis (Behcet’s Disease, Primary Angiitis of the Central Nervous System, Cogan’s Syndrome, and Erythema Elevatum Diutium) - Kenneth T. Calamia and Carlo Salvareni; F. Kawasaki’s Disease - Barry L. Myones
- **Relapsing Polychondritis** - Harvinder S. Luthra
- **Adult Onset Still’s Disease** - John M. Esdaile
- **Periodic Syndromes** - John G. Ryan and Daniel L. Kastner
- **Less Common Arthropathies - A. Hematologic and Malignant Disorders** - Adel G. Fam; B. Rheumatic Disease and Endocrinopathies - Peter A. Merkel; C. Hyperlipoproteinemia and Arthritis - Robert F. Spiera; D. Neuropathic Arthropathy - Ann K. Rosenthal; E. Dermatologic Disorders - Jeffrey P. Callen; F. Hypertrophic Osteoarthritis - Manuel Martinez-Lavin
- **Complex Regional Pain Syndrome** - Geoffrey Littlejohn
- **Sarcoidosis** - Edward S. Chen
- **Storage and Deposition Diseases** - Duncan A. Gordon
- **The Amyloidoses** - Pasha Sarraf and Jonathan Kay
- **Neoplasms of the Joint** - Andrew J. Cooper, James D. Reeves, and Sean P. Scully
- **Heritable Disorders of Connective Tissue** - Reed Edwin Pyeritz
- **Bone and Joint Dysplasias** - William A. Horton
- **Osteonecrosis** - Thorsten M. Seyler, David Marker, and Michael A. Mont
- **Paget’s Disease of Bone** - Roy D. Altman
- **Osteoporosis - A. Epidemiology and Clinical Assessment** - Kenneth G. Saag; B. Pathology and Pathophysiology - Philip Sambrook; C. Treatment of Postmenopausal Osteoporosis - Nelson B. Watts
- **Rehabilitation of Patients with Rheumatic Diseases** - Thomas D. Beardmore
- **Psychosocial Factors in Arthritis** - Alex Zautra and Denise Krukszewski
- **Self-Management Strategies** - Teresa J. Brady
- **Pain Management** - John B. Winfield
- **Therapeutic Injections of Joints and Soft Tissues** - Juan J. Canoso
- **Non-Steroidal Anti-inflammatory Drugs** - Leslie J. Crofford
- **Glucocorticoids** - Frank Buttgereit and Gerd-Rüdiger Burmester
- **Operative Treatment of Arthritis** - Joseph A. Buckwalter and W. Timothy Ballard
- **Complementary and Alternative Therapies** - Erin L. Arnold and William J. Arnold
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.
Order Now!  Primer on the Rheumatic Diseases

Yes, please send me  ____ copies

○  Primer on the Rheumatic Diseases

○ Please bill me
○ Please charge my credit card:
  ○ Eurocard/Access/Mastercard
  ○ Visa/Barclaycard/Bank/Americard
  ○ AmericanExpress

Name
Dept.
Institution
Street
City / ZIP-Code
Country
Email

Date ✖  Signature ✖

Available from

Springer Distribution Center GmbH, Haberstrasse 7, 69126 Heidelberg, Germany
Call: + 49 (0) 6221-345-4301  Fax: +49 (0) 6221-345-4229
Email: SDC-bookorder@springer.com  Web: springer.com

All € and £ prices are net prices subject to local VAT, e.g. in Germany 7% VAT for books and 19% VAT for electronic products. Pre-publication pricing: Unless otherwise stated, pre-pub prices are valid through the end of the third month following publication, and therefore are subject to change. All prices exclusive of carriage charges. Prices and other details are subject to change without notice. All errors and omissions excepted.
Primer on the Rheumatic Diseases
Klippel, J.H.; Stone, J.H.; Crofford, L.e.J.; White, P.H. (Eds.)
2008, XIX, 721 p., Softcover