Primer on the Rheumatic Diseases

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

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13th EDITION
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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- A section devoted entirely to juvenile inflammatory arthritis, with individual chapters on “Clinical Features”, “Pathology and Pathogenesis”, “Treatment and Assessment”, and “Special Considerations”.
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- A tripling of the text devoted to psoriatic arthritis, an acknowledgement of the substantial treatment advances in that disorder.
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Patients with PG also demonstrate pathergy. Thus, this condition has been reported following a variety of surgical procedures, for example, thoracotomy at heart surgery.

PG mimics other conditions, including Sweet’s syndrome, Behçet’s disease, and ankylosing spondylitis. In cases of Sweet’s syndrome, neutrophilic infiltration is accompanied by fever, erythema, and the eruption of tender nodules. In the majority of cases, there is resolution of the eruption with drug treatment; in some cases, there is persistence of lesions similar to those of Sweet’s syndrome, sometimes with a tendency toward development of pyoderma gangrenosum.

In cases of PG associated with an underlying disease (e.g., inflammatory bowel disease or RA), treatment of the latter condition often leads to improvement in PG. Pseudomon (1 mg/kg/day) is generally the first line of therapy for ulcerative PG,infliximab (5–10 mg/kg every 8 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 (300–1000 mg/day), assuming normal levels of thioctic acid and pyridoxine (1–5 g/day).

Nonsteroidal antiinflammatory drugs (NSAIDs) are contraindicated in PG because of the risk of exacerbating the disease process. Aspirin and ibuprofen are not recommended. COX-2 inhibitors are considered to be safe because of their relative lack of effect on platelet aggregation. Methotrexate, azathioprine, and leflunomide are considered to be safe in PG. However, their use may be complicated by the development of cutaneous adverse reactions.

Infectious mimickers are not common but include deep fungal infections; for example, blastomycosis, sporotrichosis, and coccidioidomycosis; as well as other conditions such as sarcoidosis, histoplasmosis, and coccidioidomycosis; as well as other conditions such as sarcoidosis, histoplasmosis, tuberculosis, and granulomatous disease of the skin. The diagnosis of PG is often difficult, and biopsy may be required to rule out other conditions.

The differential diagnosis of PG includes Sweet’s syndrome, Behçet’s disease, and ankylosing spondylitis. In cases of Sweet’s syndrome, neutrophilic infiltration is accompanied by fever, erythema, and the eruption of tender nodules. In the majority of cases, there is resolution of the eruption with drug treatment; in some cases, there is persistence of lesions similar to those of Sweet’s syndrome, sometimes with a tendency toward development of pyoderma gangrenosum.

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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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