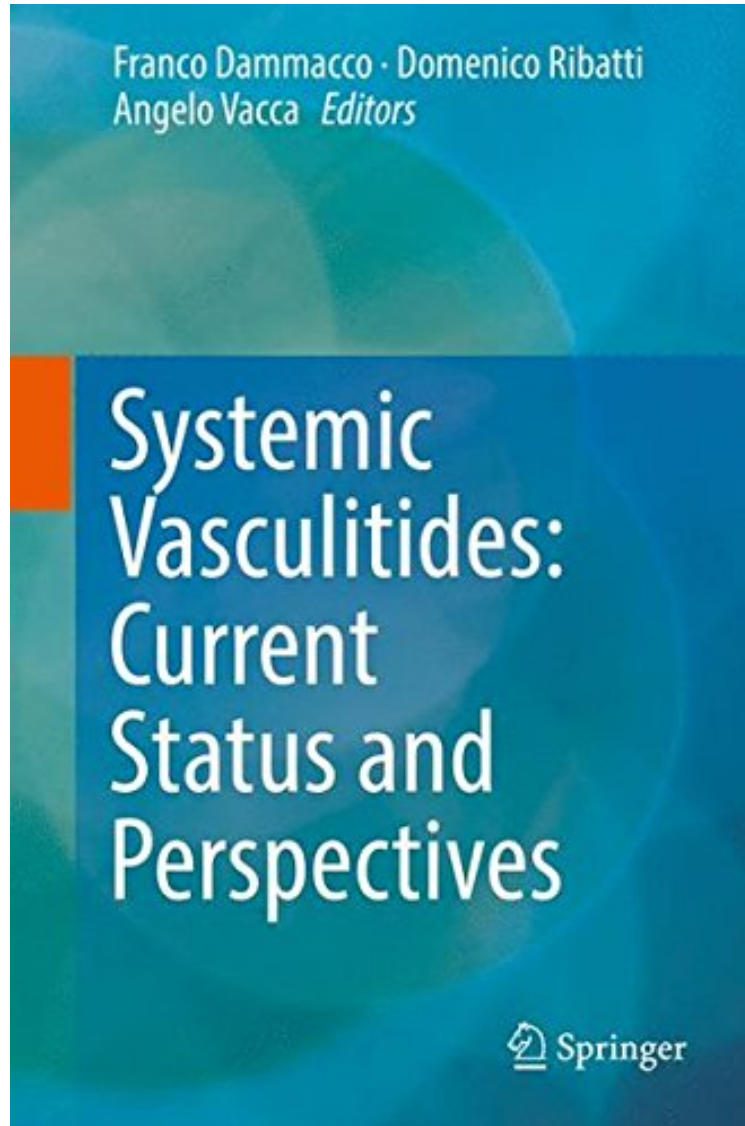


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In spite of their relatively low prevalence, systemic vasculitides have been the object of intensive basic and clinical investigations over the last few years. As a consequence, important advancements have been achieved: from updated diagnostic and classification criteria and a more rational nomenclature to the recognition of an expanding spectrum of clinical manifestations and potentially devastating complications; from the recognition of the viral etiology of conditions such as HCV-related cryoglobulinemic vasculitis and HBV-associated polyarteritis nodosa to newly named variable vessel vasculitis exemplified by Behets disease; from single-organ vasculitis such as central nervous system vasculitis to the more recently emerging features of the IgG4-related, immune-mediated diseases that are showing remarkable clinical heterogeneity. In addition, intriguing pathogenetic hypotheses are being reported for certain chronic, relapsing vasculitides that are improving our understanding of their biology and basic pathophysiology. New avenues are being explored that will hopefully allow a deeper comprehension of the relationships between certain virus-driven vasculitides and lymphoproliferation, and possibly lead to the identification of novel biomarkers that may be used to single out patients at an increased risk of relapse. This explosion of knowledge is obviously resulting in state-of-the-art, personalized treatments of systemic vasculitides. This book is a collection of reviews on the major vasculitides, written by scientists and clinicians with a multi-year experience in this field. We hope it will provide the reader with a stimulating container of new advances in scientific knowledge and more rational therapeutic approaches to this fascinating chapter of pathology.

From the Back CoverThe systemic vasculitides, including large, medium, small, and variable vessel vasculitis, have been the focus of intensive basic and clinical investigations over the last two decades. Among the important advances stemming from these efforts are new definitions, classifications, and diagnostic criteria for the different classes of vasculitis; the addition of anti-neutrophil cytoplasmic autoantibodies as a new criterion for classifying vasculitis; the recognition of the viral etiology of conditions such as cryoglobulinemic vasculitis and polyarteritis nodosa; an appreciation of the broad spectrum of clinical manifestations and potentially devastating complications associated with vasculitis; the many features and remarkable clinical heterogeneity of IgG4-related, immune-mediated diseases; and the proposal of intriguing pathogenetic hypotheses for certain chronic, relapsing vasculitides. This improved understanding of the systemic vasculitides has been accompanied by a trend away from the use of eponyms for these conditions; thus, established terms such as Wegeners granulomatosis and Churg-Strauss syndrome have been replaced by the more descriptive definitions granulomatosis with polyangiitis and eosinophilic granulomatosis with polyangiitis, respectively. Additional clinical laboratory tests, rapidly developing imaging techniques that can assess inflammation, especially in large-vessel vasculitis, and artificial neural network approaches will no doubt bring a wealth of information that ultimately leads to the identification of novel disease biomarkers. Expected applications include the identification of individuals at increased risk of relapse who would benefit from patient-tailored therapy. Although the conventional combination of glucocorticoids and immunosuppressive drugs is effective in the treatment of a large proportion of vasculitic disorders, safer medications, with fewer side effects, are being developed, including several biological agents now being closely evaluated in multi-center studies. This volume brings together comprehensive and up-to-date reviews written by experienced scientists and clinicians from many countries. Its aim is to provide readers with state-of-the-art knowledge of the major vasculitides and cutting-edge insights into their multi-faceted features. It is our hope that this book serves as a valuable and stimulating resource for basic and clinical researchers, specialists in related disciplines, as well as practicing physicians and advanced medical students interested in this fascinating branch of pathology. Franco Dammacco, Domenico Ribatti, Angelo Vacca

About the Author Franco Dammacco is Emeritus Professor of Internal Medicine at the University of Bari Medical School and has been President of the Italian Society of Internal Medicine. His research and clinical interests include HCV-related cryoglobulinemic vasculitis, onco-hematology, connective tissue diseases and immunodeficiency syndromes. He is author or co-author of more than 470 papers and Editor-in-Chief of the journal *Clinical and Experimental Medicine*. Domenico Ribatti is full Professor of Human Anatomy at the University of Bari Medical School. His main research topics are angiogenesis and anti-angiogenesis, with special emphasis for hematological malignancies, tumor microcirculation, and immunological competence. He is author of over 650 papers and 8 monographs. Angelo Vacca is full Professor of Internal Medicine at the University of Bari Medical School and President of the Italian Society of Immunology, Clinical Immunology and Allergology. He is actively involved in research areas such as angiogenesis and tumor progression, multiple myeloma and related malignancies, biological markers of tumor invasion and metastasis, and controlled chemotherapy studies. On these topics he has published over 300 papers.