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Nota di contenuto	Preface -- 1 Introduction: Nomenclature and Classification -- 2 Epidemiology -- 3 Genetics -- 4 Pathogenesis B lymphocytes T lymphocytes -- 5 ANCA: methods and clinical significance -- 6 Activity and Damage -- 7 Eosinophilic Granulomatosis with Polyangiitis (EGPA) -- 8 Granulomatosis with Polyangiitis (GPA) -- 9 Microscopic Polyangiitis (MPA) -- 10 Vasculitis /organs involvement -- 11 ENT involvement -- 12 Lung involvement -- 13 Kidney involvement -- 14 Nervous system involvement. 15 Peripheral Nervous System -- 16 Central Nervous System -- 17 Skin involvement -- 18 Miscellaneous organ/system involvement (heart, gastrointestinal, articular....) -- 19 Prognosis and outcome -- 20 Therapy -- Subject Index.
Sommario/riassunto	This volume, written by well-known experts in the field, covers all aspects of Anti-Neutrophil Cytoplasmic Antibody (ANCA) Associated Vasculitis (AAV). The expression refers to a group of diseases, characterized by destruction and inflammation of small vessels. The clinical signs vary and affect several organs, such as the kidney, lung,

skin, nervous system and others. The opening chapters give some historical hints, explain the genetic basis of the disease and provide insights into the pathogenesis derived from recent experimental studies and guides the reader through classification and nomenclature. A large part of the book is then devoted to a detailed description of the specific related diseases and their clinical presentations, the disease course, and potential complications. The advice regarding treatment is based on the best currently available evidence in this constantly evolving area. The book is part of Springer's series *Rare Diseases of the Immune System*, which presents recently acquired knowledge on pathogenesis, diagnosis, and therapy with the aim of promoting a more holistic approach to these conditions. AAVs are systemic autoimmune diseases of unknown cause that affect small (to medium) sized blood vessels. They include granulomatosis with polyangiitis (formerly Wegener's granulomatosis), microscopic polyangiitis, and eosinophilic granulomatosis with polyangiitis (formerly Churg–Strauss syndrome). This volume will be an invaluable source of up-to-date information for all practitioners involved in the care of patients with these diseases.

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