Introduction

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The connective tissue disorders (CTDs) comprise a number of related conditions that include systemic lupus erythematosus, systemic sclerosis, myositis, and Sjögren's syndrome. They are characterized by autoantibody production and other immune-mediated dysfunctions, mainly disturbances of T-cell and B-cell functions.

Basic and clinical research has been advancing rapidly in the field of CTDs, a promising world of new diagnostic tools and pharmacological agents in development that offer patients the real possibility of new therapies and physicians and scientists novel insights into the pathogenesis of these diseases.

Looking back over the past decade, research has mainly focused on the role of B cells and B-cell cytokines in the pathogenesis of CTDs and the use of anti-B-cell/anti-B-cell cytokine agents in their therapy. Looking forward, a number of new themes are emerging, including therapeutic modulation of T/B lymphocyte signaling with so-called target therapies, inhibition of T-cell activation, antibodies to IFN, IL, and anti-CD40L.

This textbook summarizes the critical aspects of the autoimmune conditions facing the clinician in the twenty-first century, including both basic science and clinical science in order to provide a translational medicine model. Two sections deal separately with systemic lupus erythematosus (SLE) and Sjögren's syndrome (SS).

SLE is a complex autoimmune disease, predominantly affecting young women during the prime years of their life. The chronic nature of the disease, its relapsing

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remitting course, and organ damage accrual over time frustrate both the physician and the patient.

To date, lupus has no known cause or cure. Early detection and treatment is the key to a better health outcome and can usually lessen the progression and severity of the disease. Anti-inflammatory drugs, antimalarials, and steroids are often used to treat lupus. Cytotoxic chemotherapies are also used to suppress the immune system in lupus patients.

Sjögren's syndrome is a systemic autoimmune disease whose clinical spectrum extends from sicca syndrome to systemic involvement (extraglandular manifestations). Systemic involvement plays a key role in the prognosis of Sjögren's syndrome, and recent studies have focused on cutaneous, pulmonary, renal, and neurological disease features. The diagnosis of Sjögren's syndrome can be confusing and time consuming. The management can also be a significant challenge for the clinician. However, recent genomic and proteomic developments are unlocking the mystery of the disease process as well as contributing to our ability to define, diagnose, and develop new treatment modalities for patients with this complex disorder.

This volume features very prominent physicians and scientists as contributors who bring their most recent discoveries to the benefit of the readers, who will find introductory contributions regarding general diagnostic and treatment principles, followed by chapters addressing the SLE and SS-specific organ manifestations. This book also offers an update on specific aspects of these diseases, including an emphasis on unifying aspects such as connections between immune system dysfunctions and development of the different types of CTDs, management of high-risk pregnancies, and the role of new target therapies.

This book would be aimed both at the rheumatologist already familiar with CTDs and at the general clinician and practitioner, equipping them to handle the requirements of the unique treatment, as well as rheumatologist trainees and nurses wishing to specialize.