1. Record Nr. UNINA9910298314503321

Titolo	Progress in Heritable Soft Connective Tissue Diseases / / edited by Jaroslava Halper
Pubbl/distr/stampa	Dordrecht : , : Springer Netherlands : , : Imprint : Springer, , 2014
ISBN	94-007-7893-7
Edizione	[1st ed. 2014.]
Descrizione fisica	1 online resource (246 p.)
Collana	Advances in Experimental Medicine and Biology, , 0065-2598 ; ; 802
Disciplina	616.77042
Soggetti	Human genetics Cell physiology Internal medicine Human Genetics Cell Physiology Internal Medicine
Lingua di pubblicazione	Inglese
Formato	Materiale a stampa
Livello bibliografico	Monografia
Note generali	Description based upon print version of record.
Nota di bibliografia	Includes bibliographical references at the end of each chapters and index.
Nota di contenuto	Structure, physiology and biochemistry of collagens -- Basic components of connective tissues and extracellular matrix -- Proteoglycans and diseases of soft tissues -- Advances in the use of growth factors for treatment of disorders of soft tissues -- Clinical, diagnostic, and therapeutic aspects of the Marfan syndrome -- Loeys-Dietz syndrome -- Connective Tissue Disorders and Cardiovascular Complications: The indomitable role of Transforming Growth Factor-beta signaling -- The Ehlers-Danlos Syndrome -- Ehlers–Danlos syndrome associated with glycosaminoglycan abnormalities -- Cutis laxa -- Collagen type VI myopathies -- Mouse Models in Tendon and Ligament Research -- Connective tissue disorders in domestic animals.
Sommario/riassunto	This volume is a reference handbook focusing on diseases like Marfan syndrome, Ehlers-Danlos syndrome, Loeys-Dietz syndrome and other heritable soft connective tissue diseases. The book presents detailed information for both basic scientists and for clinicians seeing patients. It is also a stepping stone for new investigations and studies that goes beyond the facts about the composition and biochemistry of the

connective tissue and extracellular matrix, as the authors connect individual components to specific aspects of various soft tissue disorders and to the actual or potential treatment of them. Progress in Heritable Soft Connective Tissue Diseases features very prominent physicians and scientists as contributors who bring their most recent discoveries to the benefit of readers. Their expertise will help clinicians with proper diagnosis of sometimes elusive and uncommon heritable diseases of soft connective tissues. This book also offers an update on the pathophysiology of these diseases, including an emphasis on unifying aspects such as connections between embryonic development of the different types of connective tissues and systems, and the role of TGF-beta in development and physiology of soft tissues. This new set of data explains, at least in part, why many of these disorders are interconnected, though the primary pathophysiological events, such as gene mutations, may be different for each disorder.