Animal Models for the Study of Human Disease: Chapter 41. Animal Models of Systemic Sclerosis

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Systemic sclerosis (scleroderma) is a fibrotic condition characterized by immunologic abnormalities, vascular injury, and increased accumulation of extracellular matrix (ECM) proteins in the affected organs. Although the etiology of scleroderma has not yet been fully elucidated, a growing body of evidence suggests that ECM overproduction by activated fibroblasts results from a complex interaction among endothelial cells, immunocytes, and fibroblasts, involving a number of mediators such as cytokines, chemokines, growth factors, and their receptors. For a better understanding of the pathophysiology of scleroderma, animal models are important tools. They reproduce several histological and biochemical aspects resembling human scleroderma, and we can obtain lots of new findings through animal studies. On the other hand, it must be emphasized that there are no animal models so far exhibiting all the aspects of human scleroderma, and studying animal models cannot answer all the problems of human scleroderma. This chapter introduces the current concepts of various animal models for scleroderma, and discusses their advantages/disadvantages, contribution to our understanding of the pathogenesis, and therapeutic approach for human scleroderma.

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