# REVIEW

# Renal Impairment in Systemic Sclerosis

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**Abstract**

*Systemic sclerosis (SSc) represents a connective tissue disease, characterized by progressive fibrosis of the skin and internal organs, microvascular abnormalities, and alterations in cellular and humoral immunity. Renal impairment is a relatively common feature in patients with systemic sclerosis. It can take various clinicopathological forms, of which the most specific and severe manifestation is represented by scleroderma renal crisis. This presentation is characterized by acute onset of moderate to malignant hypertension and acute kidney injury. Although some progress has been made in management of scleroderma renal crisis with the introduction of angiotensin-converting enzyme inhibitors therapy, a large population of patients still presents a poor outcome, with up to 50 percent needing renal replacement therapy. Further understanding of disease pathogenesis may lead to improvement in patient's outcome and survival.*

**Keywords:** scleroderma renal crisis, angiotensin-converting enzyme inhibitors, end-stage kidney disease.

## Introduction

Systemic sclerosis (SSc) represents a rare, chronic connective tissue disease, characterized by progressive fibrosis of the skin and internal organs, microvascular abnormalities, and alterations in cellular and humoral immunity [1].

Systemic sclerosis can be classified by clinical presentation and visceral involvement in localized scleroderma, in which the fibrosis is limited to the skin, and systemic sclerosis, which also involves the internal organs [1,2]. The spectrum of localized scleroderma includes morphea, linear scleroderma and en coup de sabre. The major subsets of systemic sclerosis include limited cutaneous systemic sclerosis (lcSSc), diffuse cutaneous systemic sclerosis (dcSSc) and systemic sclerosis sine scleroderma. The