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EM - REVIEW

Scleroderma renal crisis: a review for emergency physicians

Tim Montrief[1] · Alex Koyfman[2] · Brit Long[3]

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Abstract
Scleroderma renal crisis (SRC) remains a high-risk clinical presentation, and many patients require emergency department (ED) management for complications and stabilization. This narrative review provides an evidence-based summary of the current data for the emergency medicine evaluation and management of SRC. While SRC remains a rare clinical presentation, surveillance data suggest an overall incidence between 4 and 6% of patients with scleroderma. The diagnostic criteria for SRC include a new onset blood pressure > 150/85 mm Hg OR increase ≥ 20 mm Hg from baseline systolic blood pressure, along with a decline in renal function, defined as an increase serum creatinine of ≥ 10% and supportive features. There are many risk factors for SRC, including diffuse and rapidly progressive skin thickening, palpable tendon friction rubs, and new anemia or cardiac events. Critical patients should be evaluated in the resuscitation bay, and consultation with the nephrology team for appropriate patients improves patient outcomes.

Keywords Nephrology · Scleroderma · Renal

Introduction

Scleroderma, also called systemic sclerosis (SSc), is a rare, life-threatening, autoimmune-mediated, widespread inflammatory connective tissue condition causing fibrotic changes in the skin and vasculature, ultimately affecting major organ systems [1, 2]. The pathologic hallmark of SSc is uncontrolled accumulation of collagen and widespread vasculopathy characterized by thickening of the vascular wall and narrowing of the vascular lumen. While the exact pathogenesis of SSc remains elusive, autoantibody production, lymphocyte and fibroblast activation, vascular proliferation, obliterative microvascular disease, and connective tissue fibrosis likely play a role [3]. SSc affects women four times as often as men, with an average age of onset between 30 and 60 years of age [2, 4]. While it is believed to be associated with both genetic and environmental factors, particularly innate cellular and humoral immunity, the true etiology of SSc remains undetermined [5].

SSc is determined by the extent of cutaneous manifestations and classified as either limited cutaneous (lcSSc) or diffuse cutaneous (dcSSc) [1–4, 6]. The magnitude of skin and organ involvement directly correlates to the patient's clinical course, morbidity, and mortality, with the most dismal prognosis associated with diffuse disease [1, 2]. LcSSc predominantly affects the peripheral aspects of the body distal to the elbows and knees and is characterized by sclerodactyly and acrosclerosis [1, 7]. However, lcSSc may present with Raynaud disease, dysphagia, calcinosis cutis, telangiectasia, pulmonary hypertension, or biliary cirrhosis [7, 2].

Conversely, dcSSc involves the more proximal aspects of the extremities and the trunk, including the cardiac, pulmonary, gastrointestinal, and renal systems [1, 5–8]. Scleroderma renal crisis (SRC) is a life-threatening complication commonly associated with dcSSc, predominantly occurring within the first 5 years after disease onset [4]. The pathophysiology of SRC involves an abrupt onset of moderate to severe hypertension (> 150/85 mm Hg OR increase ≥ 20 mm Hg from baseline systolic blood pressure) over days to weeks that is typically associated with an increase in plasma renin activity and acute kidney

✉ Brit Long
brit.long@yahoo.com

[1] Department of Emergency Medicine, Jackson Memorial Hospital/Miller School of Medicine, University of Miami, 1611 N.W. 12th Avenue, Miami, FL 33136, USA

[2] Department of Emergency Medicine, The University of Texas Southwestern Medical Center, 5323 Harry Hines Boulevard, Dallas, TX 75390, USA

[3] Department of Emergency Medicine, Brooke Army Medical Center, 3841 Roger Brooke Dr, Fort Sam Houston, TX 78234, USA

injury (AKI) [1, 2, 9]. While the pathogenic mechanisms underlying SRC are not completely understood, they involve intimal thickening of the renal arteries as a result of endothelial cell injury. This results in decreased renal perfusion, with subsequent hyperplasia of the juxtaglomerular apparatus and increased renin release. Hyperreninemia causes further vasoconstriction and hypoperfusion, perpetuating the initial insult [5].

Although non-nephrotic range proteinuria, mild elevation of serum creatinine, and hypertension develop in up to 50% of patients with SSc, SRC occurs roughly in 4–6% [10, 11] of SSc patients, predominantly affecting those with dcSSc [12]. Historically, SRC has been reported to occur in up to 25% of SSc patients, whereas recent literature suggests a decrease to less than 5% of these patients and less than 2% of those with lcSSc [10]. In approximately 10% of patients, SRC occurs in the absence of hypertension, leading to the definition of normotensive SRC [4, 13]. Relative hypertension may be present, or an apparently normal blood pressure that is elevated compared to the patient's baseline values (e.g., 130/85 mmHg in a young woman whose baseline value is 100/70 mmHg).

A number of risk factors predict the occurrence of SRC, including SSc duration < 4 years, diffuse and rapidly progressive skin thickening, palpable tendon friction rubs, and new anemia or cardiac events (e.g., pericardial effusion or congestive heart failure) [14–16]. Another important risk factor for SRC is the use of glucocorticoids, particularly in high doses (e.g., prednisone > 15 mg per day), which exhibits a dose-dependent effect on the risk of SRC development [17–19]. Glucocorticoids result in salt and volume retention, the initiation or worsening of hypertension, and greater chance of SRC in a subset of patients.

Discussion

Clinical features of scleroderma renal crisis

Although there is no generally accepted or validated definition of SRC, an updated consensus classification has been proposed (Table 1), [20, 9] focusing on an abrupt onset of moderate to severe hypertension and a decline in renal function [1, 2]. Additionally, patients may present with normotensive SRC, characterized by an increase in blood pressure ≥ 20 mm Hg from baseline systolic blood pressure with a concomitant decline in renal function.

History and physical examination

The diagnosis of SRC is based on the characteristic findings in high-risk patients with SSc and primarily centers on a presentation of rapidly progressive hypertension and renal failure. The main clinical features of SRC in published data for cohorts are provided in Fig. 1. The presentation of patients with SRC, as with many diseases, may include variable history, physical examination, and diagnostic findings. Typically, patients with renal crisis do not have hypertension prior to the acute onset, and the rise in blood pressure (BP) is rapid. In cases where BP is checked regularly, normal BPs have been demonstrated as recently as 24 h prior to the diagnosis of SRC [21]. As in other causes of accelerated hypertension, patients may complain of severe headache with visual disturbances or other encephalopathic symptoms. Hypertensive encephalopathy in SRC is characterized by an acute or subacute onset of lethargy, fatigue, and confusion [22, 13]. If untreated, this hypertensive encephalopathy may lead to cerebral hemorrhage, particularly in the presence of thrombotic microangiopathy, resulting in coma and death [23]. Either focal or generalized seizures may be the first manifestation of SRC [24, 25]. Patients may present with signs and symptoms of elevated renin and accelerated hypertension, including signs of congestive cardiac failure,

Table 1 Diagnostic criteria for scleroderma renal crisis

Diagnostic criteria:
New onset blood pressure > 150/85 mm Hg OR increase ≥ 20 mm Hg from baseline systolic blood pressure
A decline in renal function, defined as an increase serum creatinine of ≥ 10%
Supportive features:
Microangiopathic hemolytic anemia
Findings consistent with accelerated hypertension on retinal examination
Microscopic hematuria on urine dipstick and/or red blood cells on urine microscopy
Oliguria or anuria
Flash pulmonary edema
Renal biopsy with typical features of scleroderma renal crisis including onion skin proliferation within the walls of the intrarenal arteries and arteriole, fibrinoid necrosis, glomerular shrinkage

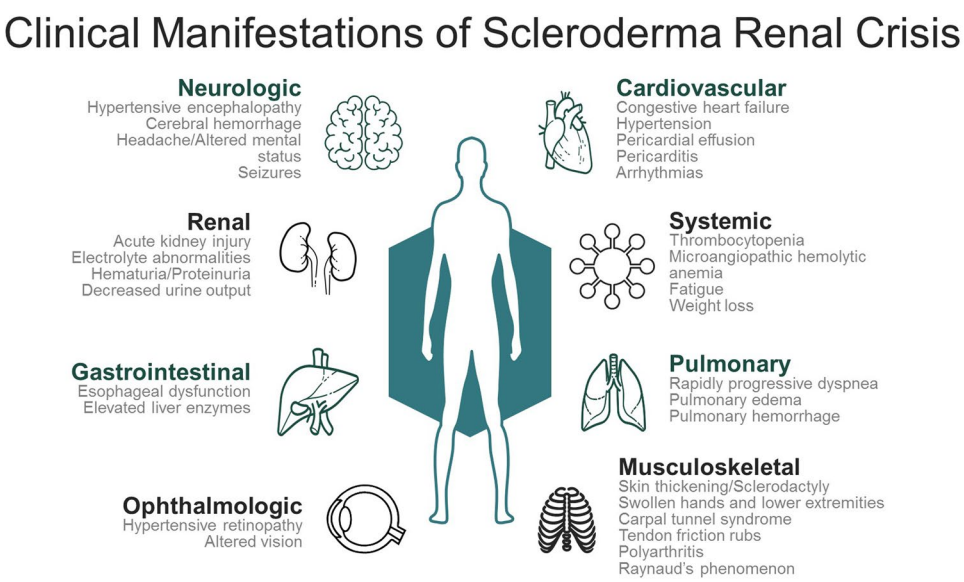

Fig. 1 Common clinical manifestations of scleroderma renal crisis

pericardial effusion, or dysrhythmias [26]. Fundoscopy may demonstrate hypertensive retinopathy [27].

The presenting BP of the patient with SRC varies, but a large majority have significant hypertension, with up to 90% having BP levels greater than 150/90 mmHg, and 30% having diastolic recordings greater than 120 mmHg [20]. A BP in the normal range is observed in approximately 10% of SRC cases, although these patients usually have a significantly raised BP compared to their baseline measurements [21].

As in other forms of AKI, patients may present with oliguria or with uremic symptoms. In severe cases of SRC, vascular occlusion and tissue ischemia may lead to renal infarcts and subcapsular hemorrhages visible on autopsy [28]. Given the rapidly progressive nature of SRC, patients may present with flash pulmonary edema due to congestive heart failure related to HTN and/or diastolic left ventricular dysfunction in the context of oliguric renal failure [20, 29]. Progressive dyspnea may present with evidence of pulmonary hemorrhage as well [13]. Approximately, half of patients with SRC will present with evidence of MAHA [5, 30]. Jaundiced or pale skin, dark urine, and splenomegaly are found in patients with MAHA.

Thus, in any patient presenting with malignant hypertension or AKI, SSc should be considered. Clinical features that help identify patients with SSc in this context are recent-onset Raynaud's phenomenon, acute onset of fatigue, weight loss, polyarthritis, swollen extremities, carpal tunnel syndrome, and tendon friction rubs [31, 32]. The skin thickening that progresses to a diffuse form of SSc usually presents after a few months from the first symptoms [22]. However, it is important to note that SRC can occur in patients without evidence of skin thickening or other manifestations of SSc [33, 34].

Differential diagnosis of SRC

Discovering the underlying etiology of acute renal failure as a complication of SSc is not always obvious, and the diagnosis of SRC is challenging. In case of acute renal failure with SSc, a number of diagnoses should be considered (Table 2) [35]. Renal arterial stenosis can present with malignant HTN [36, 37]. Hypovolemia can mimic SRC. Thrombotic thrombocytopenic purpura (TTP), anti-neutrophil cytoplasmic antibody (ANCA)-associated glomerulonephritis, crescentic rapidly progressive glomerulonephritis (RPGN), and atypical hemolytic uremic syndrome (aHUS), which remain uncommon presentations of acute renal failure in SSc, can present similarly to SRC [38–40]. Differentiating these conditions is crucial

Table 2 Differential diagnosis for Scleroderma Renal Crisis

Renal arterial stenosis
Thrombotic thrombocytopenic purpura (TTP)
Anti-neutrophil cytoplasmic antibody (ANCA)-associated glomerulonephritis
Crescentic rapidly progressive glomerulonephritis (RPGN)
Atypical hemolytic uremic syndrome (aHUS)
Pauci-immune ANCA-associated vasculitis (AAV) with glomerulonephritis

to enable effective management and prognostication for these patients.

Even in a patient with cutaneous signs of SSc, the presence of microangiopathic hemolytic anemia and accelerated hypertension or the findings of thrombotic microangiopathy raise the possibility of a primary hematological diagnosis, including thrombotic thrombocytopenic purpura and atypical hemolytic uremic syndrome [38]. A low ADAMTS13 activity is a key feature of TTP, which helps distinguishing it from SRC-related thrombotic microangiopathy, although a renal biopsy might be needed to confirm this diagnosis [20, 41, 42]. In most institutions, results of the ADAMTS13 assay will not be available early enough to affect the immediate clinical management. Furthermore, there have been reports of patients presenting with both scleroderma renal crisis and thrombocytopenic purpura [43–45]. The distinction has significant clinical importance as plasmapheresis, the primary treatment for TTP or atypical HUS in the acute phase, is not an effective treatment for patients with SRC. Fever and hemorrhagic manifestations are the principal clinical findings that differentiate cases of thrombocytopenic purpura and atypical HUS from SRC [20].

Pauci-immune ANCA-associated vasculitis (AAV) with glomerulonephritis is another potential cause of acute renal failure in SSc patients [46–50]. However, AAV is most likely to be seen late in the disease course in patients with limited cutaneous systemic sclerosis than in patients with other forms [51]. Likewise, less than 1% of patients with SSc develops ANCA-associated vasculitis, although up to 12% of these patients have ANCA antibodies [52, 53]. It is important to note that malignant HTN and thrombotic microangiopathy are often absent in cases of AAV with glomerulonephritis [53–55]. Distinguishing between ANCA-associated vasculitis with glomerulonephritis and SRC is important, as treatment and prognosis differ greatly between the two. For instance, intravenous or oral administration of cyclophosphamide and rituximab induce and maintain remission in most patients with ANCA-associated vasculitis and glomerulonephritis; however, this is not a demonstrated treatment for scleroderma renal crisis [56].

A number of substances may precipitate SRC, including cocaine and cyclosporine [57, 58]. Additionally, thrombotic microangiopathy, characterized by hemolytic anemia and thrombocytopenia, occurs in up to 50% of these cases [22, 18]. Similarly, patients presenting with proteinuria in the nephrotic range may be due to NSAID toxicity, and any intentional or unintentional ingestions should be considered [17, 57].

Laboratory and imaging considerations

When evaluating a patient for SRC, laboratory assessment and imaging are guided by the presentation and differential diagnosis. It is reasonable to obtain a complete blood count and serum chemistries evaluating for anemia, thrombocytopenia, electrolyte abnormalities, and kidney function. A urinalysis with microscopy and peripheral blood smear may help further characterize AKI and thrombocytopenia. Laboratory findings consistent with SRC include a markedly elevated serum creatinine (increased $\geq$ 10% from baseline). Urinalysis frequently shows mild proteinuria (0.5 to 2.5 g/l), and microscopic hematuria, which corresponds to hemoglobinuria in most cases [18, 22]. Additional investigations may include coagulation studies, including fibrinogen, and cardiac biomarkers. Rheumatologic testing, while not required, may assist in differentiating SRC from other rheumatological disorders; although due to the long turn-around time for results, these may be impractical in the acute care setting but are helpful for the inpatient team. Antinuclear antibodies (abs) are common in patients with SRC, including anti-topoisomerase abs, anti-RNA polymerase III abs, and anti-centromere abs [59]. When distinguishing SRC from TTP, ADAMTS13 activity may assist, although it is rarely available in the ED.

Imaging is not necessary to diagnose SRC, although it can provide valuable information. Renal ultrasound is typically unremarkable in SRC but may be useful in some patients to rule out urinary tract obstruction or nephrolithiasis as a cause of AKI. Additionally, the renal vascular resistive index, a measure of intrarenal vascular elasticity and compliance assessed using Doppler ultrasound, can be helpful. The resistive index is sensitive to renal vascular disease and correlates with GFR and digital microvascular damage in scleroderma [60]. In the patient presenting with acute pulmonary edema, point-of-care echocardiography may show pericardial effusions and left ventricular systolic dysfunction, which are common findings secondary to the increased afterload on the heart in SRC. Signs of pulmonary hypertension are occasionally seen on echocardiogram, but in patients with SRC, this is primarily a transient secondary phenomenon due to accelerated hypertension rather than chronic pulmonary arterial hypertension [19, 20]. A chest x-ray may exclude other causes of acute dyspnea, including pneumonia and pneumothorax, and a computed tomography scan of the brain can be evaluated for intracranial catastrophes in the encephalopathic patient.

Management of scleroderma renal crisis

General considerations

If left untreated, SRC can progress to end-stage renal disease (ESRD) over a period of 1–2 months, with death usually

occurring within 1 year [21]. While the early diagnosis and initiation of treatment in SRC remain difficult, a prompt recognition of SRC is imperative. The mainstay of therapy in SRC is effective and prompt BP control, which is demonstrated to improve or stabilize renal function in approximately 70% of patients and improve survival to nearly 80% at 1 year [61]. However, the success with antihypertensive therapy is dependent on its initiation before irreversible renal damage has occurred [62]. In addition to close hemodynamic monitoring and renal replacement therapy (RRT) when required, a higher level care of the patient with SRC may include ventilatory support in the patient with severe pulmonary edema and sedation or anti-seizure medications for those with hypertensive encephalopathy. Early consultation with both nephrology and intensive care teams, when appropriate, should be sought. Given the remaining diagnostic uncertainty in this field, if a diagnosis of TTP or HUS is suspected in a scleroderma patient, plasmapheresis should be considered in close consultation with a hematologist [38].

A methodical reduction in BP is recommended, as a precipitous decrease leads to reduced renal perfusion and increases the risk of acute tubular necrosis. The eventual goal is to reach the patient's pre-SRC BP with 72 h [63]. In the absence of a diagnosis of hypertensive emergency, a steady reduction in the systolic blood pressure (SBP) of 20 mm Hg and diastolic blood pressure (DBP) of 10 mm Hg per day is preferable [20]. Although SRC-related hypertension is acute, rapid BP reduction to baseline has not been shown to bear the same risks as seen with rapid BP reduction in patients with chronic hypertension; nevertheless, conventional practice standards are not to exceed a maximum reduction in the SBP of 20 mm Hg and DBP of 10 mm Hg per day. However, in the case of hypertensive emergency, aim to reduce the mean arterial pressure by 10–20% within 1 h, with a goal DBP of 100–110 mm Hg within 24 h [64].

ACE inhibitors (ACEi)

The optimal antihypertensive agent for SRC is an ACEi. The underlying pathophysiology includes decreased renal perfusion, with subsequent hyperplasia of the juxtaglomerular apparatus and increased renin release. Hyperreninemia causes further vasoconstriction and hypoperfusion, perpetuating the initial insult [5]. An ACEi breaks this viscous cycle by disrupting the renin–angiotensin–aldosterone system.

If there is a high degree of clinical suspicion for SRC, an ACEi should be introduced or the dose increased if the patient is already taking one at home [65, 63]. A short-acting ACEi (e.g., captopril) may theoretically be preferable in the hemodynamically unstable patient, but there is little evidence that it is preferable in general to a once-daily medication (e.g., Enalapril or Ramipril) [62]. A short-acting ACEi has the advantage of rapid onset with peak effect at 60–90 min and short duration of action, permitting rapid dose titration compared with enalapril, which is not routinely used in the ED due to its longer duration of action (up to 36 h) [66, 20]. In consultation with a nephrologist, a long-acting ACEi may be added at a moderate dose (e.g., Ramipril 5 mg) [20].

Among hypertensive patients without evidence of central nervous system involvement (e.g., encephalopathy, papilledema), captopril is begun at a dose of 6.25–12.5 mg with a progressive dose escalation in 12.5–25 mg increments at 4–8-h intervals until the goal BP is reached [5]. The maximum captopril dose is 300–450 mg/day. For hypertensive patients with evidence of central nervous system involvement, we administer the same captopril dose escalation regimen and, for further acute BP control, add intravenous agents. In normotensive SRC patients, we initiate captopril at a dose of 6.25 mg, and if tolerated, increase the dose to 12.5 mg at the second dose [5]. Further dose escalation must be accomplished carefully to prevent hypotension, with titration in the inpatient setting. For those patients who have BPs within the normal range, yet still higher than the patient's baseline, the goal lowers the BP to the previous baseline.

ACEi resistance is more typical than oversensitivity in this population, and the general standard of care is to initiate a long-acting drug as soon as possible and escalate the dose to the daily maximum, although this may be deferred to the inpatient setting [5, 20]. Any rise in serum creatinine after initiating an ACEi should not trigger dose reduction or ACEi cessation, as the rise in serum creatinine is likely secondary to the underlying SRC rather than the ACEi [20]. There is no evidence in SRC that renal function can be spared or improved by minimizing ACEi dose.

Other BP lowering agents

While theoretically angiotensin receptor blockers (ARBs) should prove effective in SRC, these agents have not been adequately evaluated in this setting, and efficacy is not established [67, 68]. However, due to the limited evidence, consensus opinion recommends these agents as possible second-line agents if hypertension is unresponsive to an ACEi (Fig. 2) [20, 69]. There is no evidence regarding the role of direct renin inhibitors [22]. Dihydropyridine calcium channel blockers (CCBs), most commonly nifedipine, are appropriate for the treatment of vasospastic conditions, including Raynaud disease, which occurs in more than 90% of dcSSc patients. CCBs, particularly short-acting, are commonly recommended agents for SRC resistant to ACEi and/or ARBs (Table 3) [69, 63]. Other antihypertensive drugs that can be added to ACEi monotherapy, if necessary, include diuretics for volume overload, and/or centrally acting α-blockers such as clonidine [63, 69]. Although α-blockers may increase the

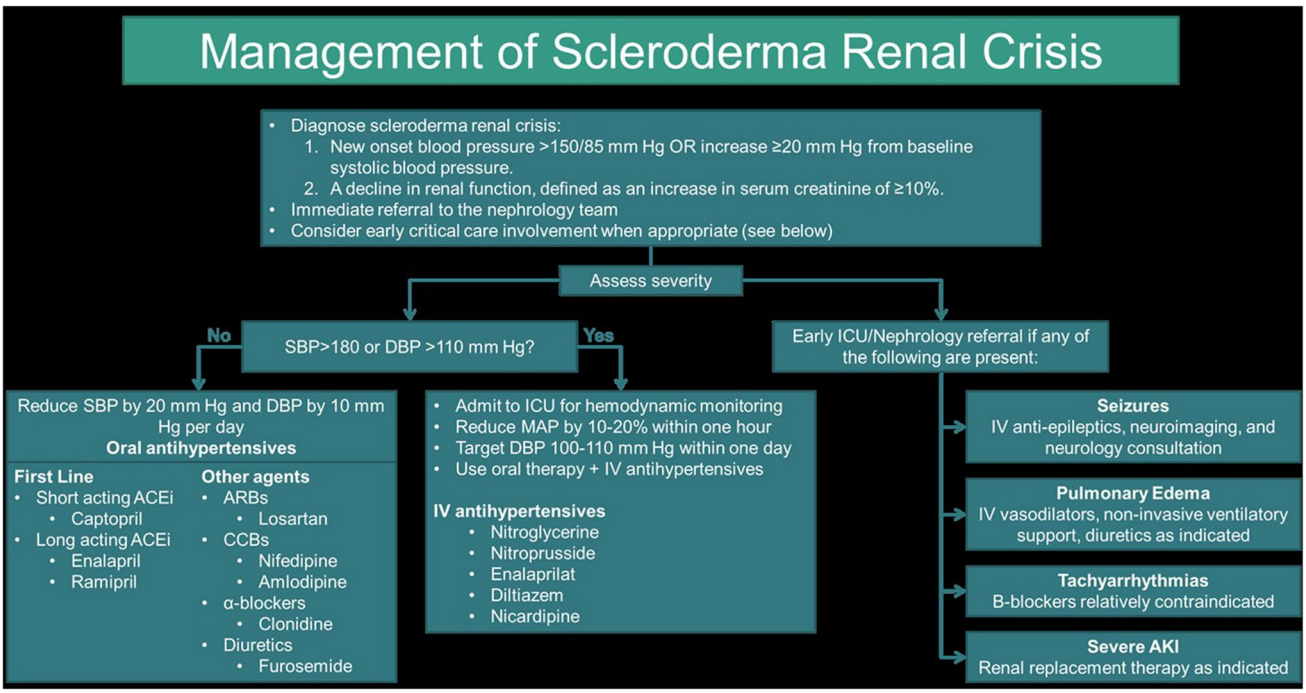

Fig. 2 Management of scleroderma renal crisis. *SBP* systolic blood pressure, *DBP* diastolic blood pressure, *ARs* Angiotensin receptor II blockers, *CCBs* calcium channel blockers. Adapted from Lynch BM, Stern EP, Ong V, Harber M, Burns A, Denton CP. UK Scleroderma Study Group (UKSSG) guidelines on the diagnosis and management of scleroderma renal crisis. Clin Exp Rheumatol. 2016;34 Suppl 100(5):106–109

likelihood of hypotension when used in combination with an ACEi, expert consensus recommends these agents as adjunctive treatments [63, 70]. Beta-blockers are usually avoided in patients with SSc due to the theoretical risk of worsening vasospasm, including Raynaud phenomenon [71]. The addition of endothelin-1 receptor antagonists (e.g., bosentan) has been used in patients with resistant hypertension. However, the long-term safety of these agents has not been demonstrated and they are not routinely suggested as pharmacotherapy for SRC [72, 73].

In patients presenting with signs and symptoms of hypertensive emergency, more aggressive management may be pursued [63]. The parenteral antihypertensive agents most often used in the initial treatment of these patients with SRC include nitroglycerin, clevidipine, sodium nitroprusside, and enalaprilat [65, 64, 74, 75]. These infusions should be discontinued as soon as possible while increasing the dose of the short-acting ACEi.

Dialysis and renal transplantation

Despite appropriate ACEi therapy, dialysis is needed in approximately 60% of patients with SRC [12, 22]. If indicated, either hemodialysis or continuous peritoneal dialysis is an effective therapy for ESRD due to SRC [5, 61, 76, 77]. There is limited experience with regard to renal transplantation in patients with SRC, in part because transplantation is sometimes precluded by the severity of the extrarenal manifestations of SSc. Historically, expert consensus was to consider renal transplant in those patients requiring dialysis who do not recover kidney function within 2 years; however, this is controversial, and some experts argue for emergent kidney transplant in patients with new-onset ESRD [5, 63, 78]. Early consultation with a nephrologist is encouraged for all cases of SRC.

Alternative therapies

Historically, treatment with the copper chelating agent d-penicillamine was once believed to be clinically beneficial, but has fallen out of favor in contemporary literature [79, 80]. Recent literature demonstrates d-penicillamine to be associated with significant adverse effects with no reduction in morbidity or mortality. Any benefit from d-penicillamine must be balanced with the risk for bone marrow suppression, further renal injury, gastrointestinal intolerance, and dermatologic complications such as pemphigus vulgaris [63, 2]. Therefore, its use is strongly discouraged in patients with SRC [61, 63].

Table 3 Antihypertensive agents in scleroderma renal crisis

Class	Drug	Dosing	Comments
ACEi	Captopril	Initial: 6.25 to 12.5 mg with a progressive dose escalation in 12.5 to 25 mg increments at 4- to 8-h intervals Maximum: 300 to 450 mg/day	First line agent Short-acting
	Enalapril	Initial: 5 mg once daily Maximum: 40 mg/day	First line agent Long acting
	Ramipril	Initial: 2.5 mg once daily Maximum: 20 mg/day	First line agent Long acting
	Enalaprilat	Initial: 1.25 mg IV injection over 5 min, administered every 6 h Maximum: 20 mg/day	Intravenous medication
ARB	Losartan	Initial: 50 mg once daily Maximum: 200 mg/day	Alternate agent
Diuretic	Furosemide	Initial: 40 mg IV once, if response not adequate within 1 h, may increase to 80 mg/dose Maximum: 200 mg/dose	Alternate agent
α-blocker	Clonidine	Initial: 0.1 mg twice daily Maximum: 2.4 mg/day	Alternate agent
CCB	Nifedipine	Initial: 30 or 60 mg once daily, extended release Maximum: 180 mg/day	Alternate agent
	Amlodipine	Initial: 5 to 10 mg once daily Maximum: 20 mg/day	Alternate agent
	Nicardipine	Start: 2.5–5 mg/h Titrate: 2.5 mg/h every five minutes Max: 15 mg/h	Intravenous infusion
	Diltiazem	Bolus: 0.25 mg/kg IV bolus over two minutes; in 15 min may repeat 0.35 mg/kg IV bolus over two minutes Infusion: 5–15 mg/h	Intravenous infusion
	Clevidipine	Start: 1–2 mg/h Titrate: Double the dose at 90-second intervals, and then increasing the dose by less than double at intervals of 5–10 min Max: 16 mg/h	Intravenous infusion
IV nitrate	Nitroprusside	Start: 0.25–0.3 mcg/kg/minute Titrate: 0.5 mcg/kg/minute every two minutes Max: 10 mcg/kg/minute	Intravenous infusion
	Nitroglycerin	Start: 5 mcg/minute Titrate: 5 mcg/minute every three to five minutes to response; if no response seen at 20 mcg/minute, may titrate up by 10–20 mcg/minute Max: 400 mcg/minute	Intravenous infusion

ACEi ACE inhibitors, *ARB* angiotensin II receptor blocker, *CCB* calcium channel blocker

Therapies for SSc

Treatment regimens for SSc without evidence of SRC are aimed at improving peripheral circulation, preventing the synthesis and release of harmful cytokines, and inhibiting fibrosis [8]. This is commonly accomplished using immunosuppressive agents, including vitamin D analogues, UV-A phototherapy, corticosteroids, cyclosporine, azathioprine, and methotrexate [2, 81].

Prognosis and disposition

Before the 1970s and the widespread use of ACEi's, SRC frequently resulted in renal failure and death, usually within months of diagnosis [61]. The use of an ACEi greatly improves the prognosis of SRC, with current patient survival of 70–82% at one year, which decreases to 50–70% at 5 years for those requiring continued dialysis [73]. However, patients who survive SRC without the need for dialysis or who only require temporary dialysis have excellent outcomes with a 5-year survival of 90% [63]. Unfortunately, there has been no clear trend towards improvement in these measures over the past 30 years. Risk factors for mortality in these patients include male sex, older age, lower BP at the time of diagnosis, and the development of congestive heart failure [18, 61, 62, 82]. Many of these patients will require close hemodynamic monitoring only available in the intensive care setting.

Conclusions

SRC remains a rare diagnosis, affecting up to 6% of patients with SSc, but continues to have a large burden of morbidity and mortality. Due to the fact that the presentation of SRC is variable, with some patients being hypertensive and others being normotensive, and some with evidence of renal insufficiency, clinicians should be aware of potential presentations and clinical histories associated with SRC. Prompt recognition and initiation of aggressive antihypertensive therapy with an ACEi in the ED may improve patient outcomes. Early consultation with the critical care and nephrology teams is important, as roughly 60% of SRC patients will require dialysis.

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Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Statement of human and animal rights This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent None.

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