Scleroderma Renal Crisis

Malignant hypertension with acute kidney injury in systemic sclerosis · an ACE inhibitor is the treatment, and steroids are the trigger

Captopril
and do not stop it

WHO IS AT RISK *this is largely a predictable complication, so know who to warn*

DISEASE FACTORS

Early diffuse cutaneous systemic sclerosis, typically within the **first 4 years** and often while the skin is still thickening rapidly. **Anti-RNA polymerase III antibody carries the highest risk** of all, so its presence should change how closely you monitor. Uncommon in limited cutaneous disease.

⚠ DRUG TRIGGERS

Glucocorticoids are the classic precipitant, and the risk rises with dose, so **steroids should be avoided or kept to the lowest possible dose in early diffuse disease**. **Cyclosporine** is also implicated. If steroids are unavoidable, **monitor blood pressure and creatinine closely** and tell the patient what to watch for.

WHAT PATIENTS SHOULD DO

At-risk patients should **monitor their own blood pressure at home** and be told to seek urgent care for a **new rise**, headache or visual change. Because the crisis moves in days, **self-monitoring is the intervention that buys the kidney**.

RECOGNIZE IT *and the mimic that leads people astray*

Feature	Detail
Abrupt hypertension	Often **malignant**, with headache, visual disturbance, encephalopathy, seizures or **PRES**, flash pulmonary edema, and retinopathy. **A patient with systemic sclerosis whose blood pressure suddenly rises has renal crisis until proven otherwise**
Acute kidney injury	Rapidly rising creatinine, with **bland urine or only mild proteinuria**. Heavy proteinuria or an active sediment should make you look for another cause
Microangiopathic picture the mimic	**Schistocytes, hemolysis and thrombocytopenia**, because this is a thrombotic microangiopathy of the renal arterioles. **It looks like TTP or HUS**, and the distinguishing feature is the systemic sclerosis and the accompanying hypertension. **The treatment is an ACE inhibitor, not plasma exchange**, so making this distinction changes everything
Normotensive crisis	**Roughly 10% have a normal blood pressure**, most often those already on steroids. **Do not exclude the diagnosis on a normal reading**: compare with the patient's usual pressure, since a rise from 90 to 130 systolic is a large change for them. Normotensive crisis carries a worse outcome

THE ANTIBODY PREDICTS THE ORGAN *which is why it is worth sending early*

Antibody	Usual subtype	The complication it points to
Anti-RNA polymerase III	**Diffuse cutaneous**, often with rapidly progressive skin thickening	**Renal crisis**, the highest-risk group. Also associated with **malignancy around the time of onset**, so an age-appropriate cancer review is reasonable
Anti-Scl-70 *anti-topoisomerase I*	**Diffuse cutaneous**	**Interstitial lung disease**, which is now the leading cause of death in systemic sclerosis. Screen with high-resolution CT and pulmonary function tests
Anti-centromere	**Limited cutaneous** (the CREST pattern)	**Pulmonary arterial hypertension**, typically later in the disease. **Annual echocardiography**, and remember PAH can occur with normal lung parenchyma

Limited versus diffuse is defined by how much skin is involved, proximal to the elbows and knees or not, and it predicts which complications to watch for rather than how severe the disease feels. **Renal crisis clusters in diffuse disease, pulmonary hypertension in limited disease**, so the subtype should shape your monitoring from the first visit.

TREATMENT *the whole management rests on one drug class*

Action	Detail
Start an ACE inhibitor immediately	**Captopril is preferred** because it is short acting and can be titrated rapidly, which suits a patient whose pressure is moving. Start as soon as the diagnosis is suspected and **uptitrate aggressively**, aiming to bring systolic down in a controlled way over about 72 hours rather than instantly
⚠ Keep going even as creatinine rises	**The single most important instruction on this sheet.** The creatinine will often rise after starting, and that is expected rather than a signal to stop. **Stopping the ACE inhibitor for a rising creatinine converts a recoverable crisis into permanent dialysis dependence.** Continue it even if dialysis is needed
⚠ ARBs are not a substitute	The evidence for benefit is with **ACE inhibitors specifically**. Do not swap to an ARB in acute crisis for a cough or convenience. Other agents can be **added** for residual hypertension, but the ACE inhibitor is never the one you drop
Dialysis is often temporary	Around half of those who need dialysis eventually come off it, and **recovery can take many months, sometimes up to 2 years**. **Do not list for transplant early**, and continue the ACE inhibitor while on dialysis to allow recovery
Then look for the rest of the disease	Screen for the other organ involvement that determines prognosis: **ILD** (high-resolution CT and pulmonary function tests), **pulmonary arterial hypertension** (annual echo), plus GAVE, reflux, dysmotility and digital ulcers

THE PROPHYLAXIS PARADOX & PEARLS

- **Do not use ACE inhibitors to prevent renal crisis.** This is counterintuitive given they treat it, but cohort data have associated **prior ACE inhibitor use with a higher risk of subsequently developing renal crisis**, and possibly with worse outcomes when it occurs. **Treat established crisis with them, and do not prescribe them prophylactically** in systemic sclerosis purely on risk.
- **Check for anti-RNA polymerase III in diffuse disease.** A positive result identifies the highest-risk group and justifies home blood pressure monitoring and extra caution with any steroid.
- **Weigh steroids explicitly against this risk.** When a patient with early diffuse systemic sclerosis needs steroids for myositis, arthritis or ILD, use the **lowest effective dose for the shortest time**, and document that the crisis risk was considered and monitoring arranged.
- **Renal crisis is now uncommon but still lethal when missed**, and its outcome depends almost entirely on **how quickly the ACE inhibitor is started**. A day of delay while a microangiopathy workup is completed is a day of nephrons lost.

References: EUSTAR analyses of scleroderma renal crisis risk factors · published cohort and review literature on ACE inhibitors in renal crisis · EULAR recommendations for the treatment of systemic sclerosis.

Educational aid; verify against current guidance, local formulary and patient factors.

roundsrx.com
roundsrx Infographic Series · #165