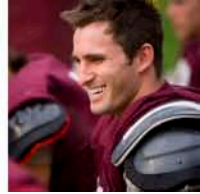




# Scleroderma Renal Crisis

**Michael Walsh, MD MSc FRCPC**  
**Departments of Medicine and Clinical Epidemiology &**  
**Biostatistics**  
**McMaster University**  
**Nephrologist**  
**St. Joseph's Healthcare**



# Conflicts of Interest

- Nothing to declare

# Objectives

- By the end of this presentation you should be able to answer the following questions:
  - Why is a scleroderma renal crisis (SRC) bad?
  - Who is likely to get it?
  - How can we prevent it and detect it?
  - What do we do once it is detected?
  - When should you refer a patient with systemic sclerosis (SSc) to a nephrologist?

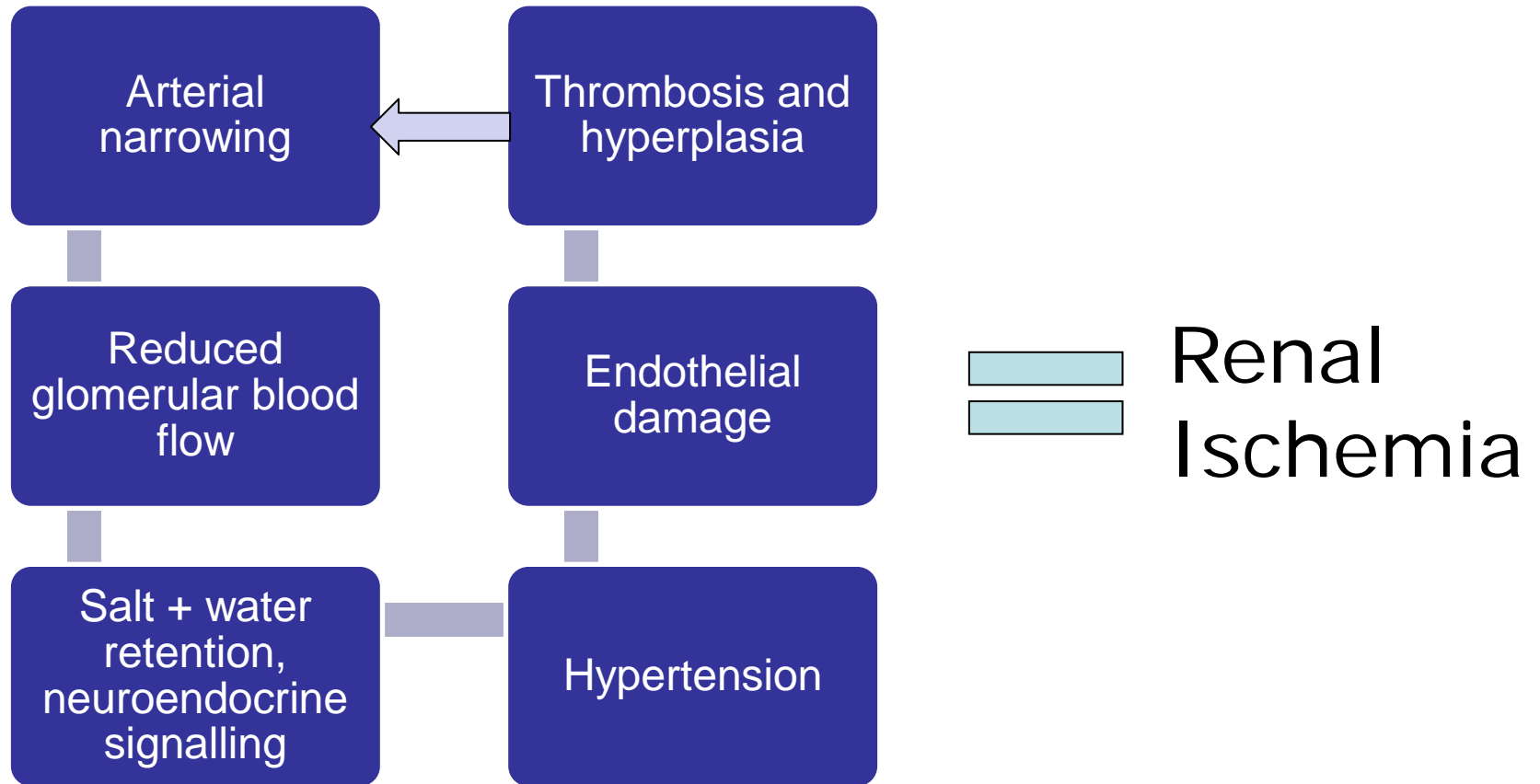
Scleroderma renal crisis

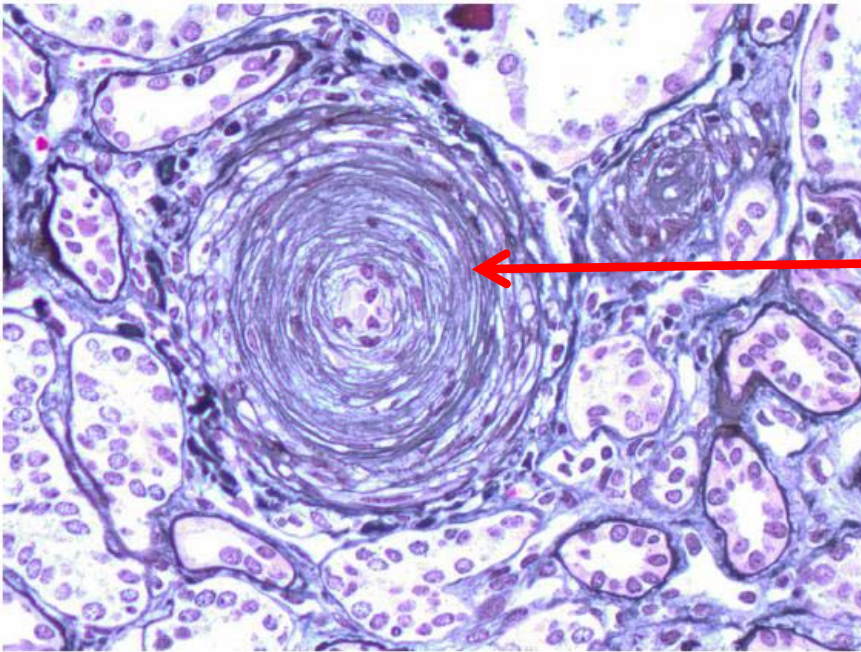
**WHY IS A SRC BAD?**

# What is a scleroderma renal crisis?

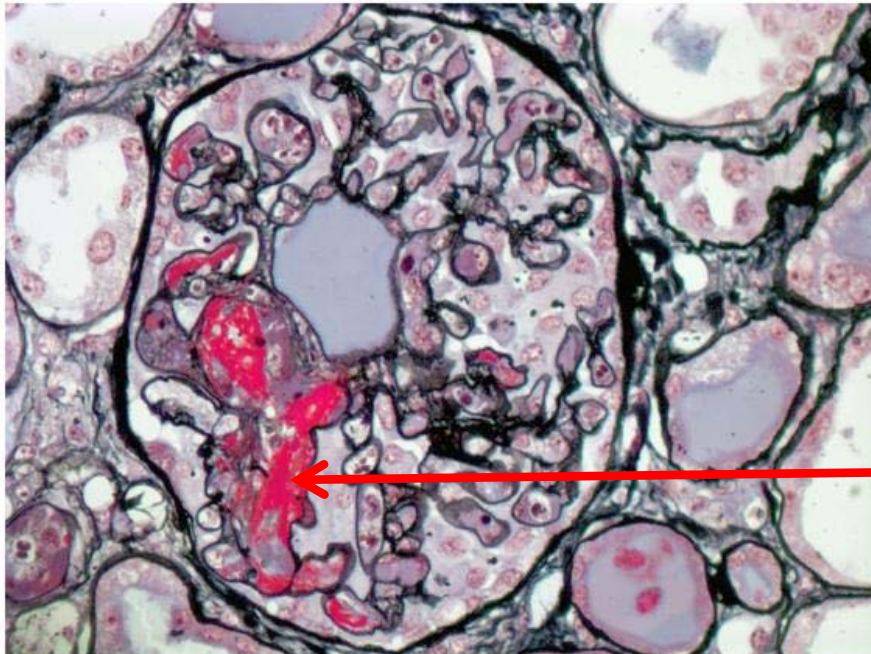
- Progressive renal failure with a (mostly) normal urine sediment
- Abrupt onset of hypertension
  - 10% normotensive

# What is a scleroderma renal crisis?





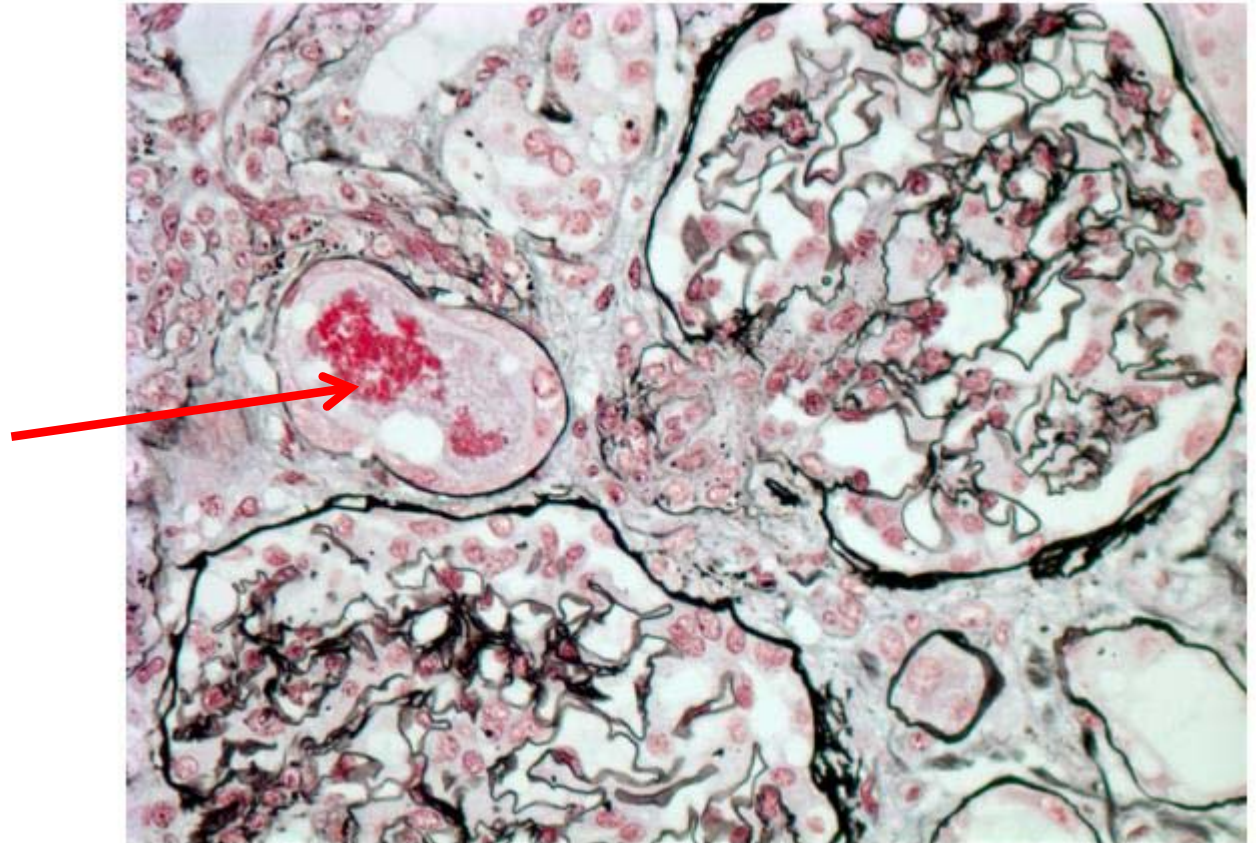
Onion skinning



Glomerular  
capillary  
thrombosis



Renin about  
to explode  
out of the  
JGA!



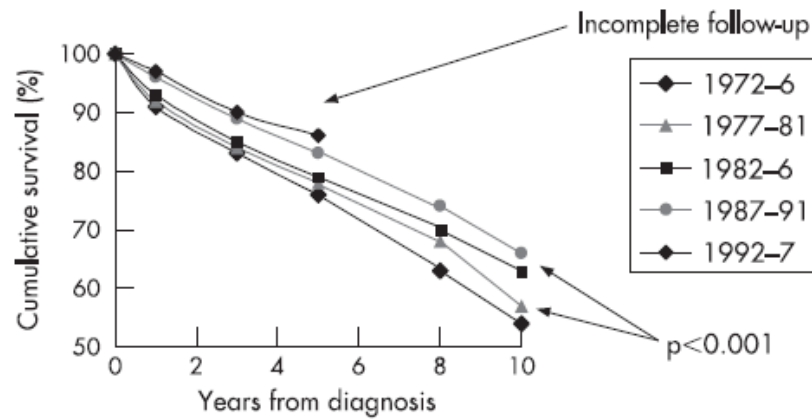


# Why is SRC bad?

- ~25% mortality in 1 year
- 20 to 50% risk of developing end-stage renal disease
  - 2 year mortality 50% (vs 36% in non-scleroderma dialysis)

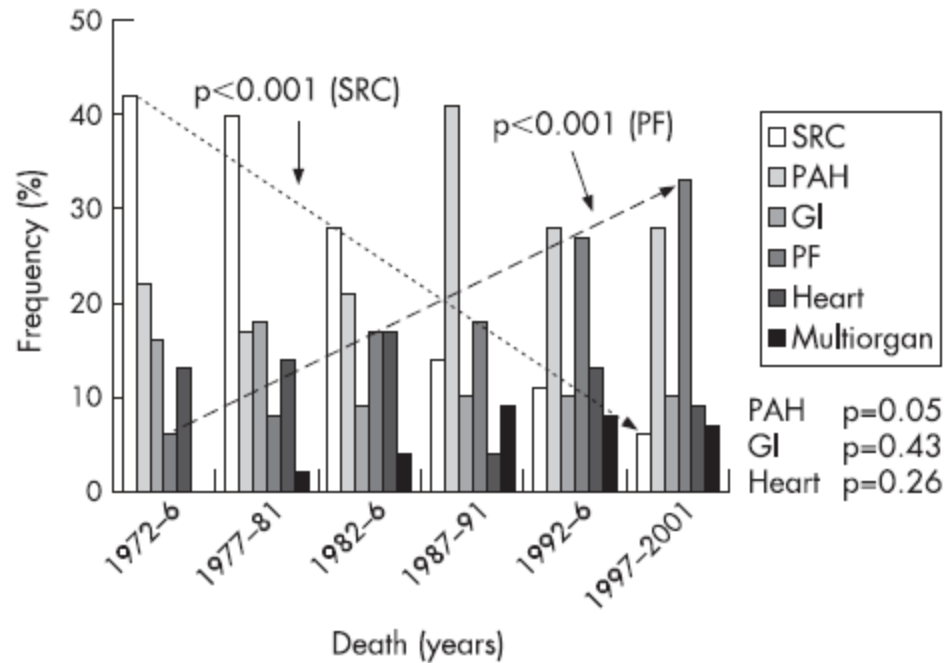


Steen et al. J Rheum. 2005.  
Steen et al. Arthritis Rheum. 1998.  
Abbott et al. JNephrol. 2002.



**Figure 1** Survival of patients with systemic sclerosis between 1972 and

### Causes of death in scleroderma



**Figure 2** Changes in causes of systemic sclerosis-related deaths between 1972 and 2001. GI, gastrointestinal; PAH, pulmonary arterial hypertension; PF, pulmonary fibrosis; SRC, scleroderma renal crisis.

- Death rate falling
- SRC as cause of death falling

Scleroderma renal crisis

**WHO GETS SRC?**

# Who gets SRC?

- 60-80% of patients with SSc have renal disease
  - 50% have proteinuria, CKD, and/or hypertension
- 20% with diffuse cutaneous SSc get SRC
  - Uncommon in patients without limited disease
  - Can happen in sine scleroderma

# Risk Factors

- Recent onset, diffuse SSc (80%)
  - 50% in first year
  - Most in first two years
- Active, progressive disease
  - 2-fold increase risk
- $\geq 15$  mg/day prednisone use
  - 4-fold increase risk
- Presence of anti-RNA polymerase III
  - ~50% SRC vs 10% nonSRC
- Other evidence of severe disease
  - Anemia (MAHA), tendon friction rubs, recent cardiac events

# Warning Bells!

- Blood pressure
  - New onset hypertension (10% normotensive)
  - Encephalopathy/seizures/retinopathy
- Kidney function
  - Creatinine rising (eGFR falling)
  - Pulmonary edema



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**HOW CAN WE PREVENT AND  
DETECT SRC?**

# Preventing SRC

- Avoid risk factors
  - Avoid glucocorticoids
    - Minimize dose and duration
  - Avoid cyclosporine
- ? Avoid prophylactic ACEi
  - Unclear evidence
- Detect early
  - Time is kidney!
  - Regular serum creatinine and proteinuria
    - Q3 months with additional for symptoms
  - Blood pressure monitoring
    - Home monitoring ideal

# Detection

- Normotensive patients
  - Rise of SBP by >20 mmHg
  - Rise of DBP by >10 mmHg
- Chronically hypertensive patients
  - >150/90 at least twice in 24 hrs despite treatment
- New proteinuria
- Sustained rise in creatinine
  - Measure at least twice
- Renal biopsy
  - When not clinically obvious

# Differential

- Any other cause of Acute Renal Failure
  - Pre-renal – rehydrate them if clinically indicated even if BP high (pressure natriuresis)
  - Post-renal
  - Renal
    - Medications (NSAIDs, antibiotics, penicillamine)
    - GN (TTP/HUS, malignant hypertension, ANCA, lupus overlap)
- Chronic?

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# **TREATMENT OF SRC**

# Treatment of SRC

- Blood pressure control
- ACEi favoured
  - Captopril favoured
    - Rapid onset
    - Short duration of action
    - Easy to titrate
    - Can use very high doses (e.g. 50 q4h)
  - Associated with more kidney recovery and lower mortality
    - Low study quality
  - Monitor creatinine and potassium carefully!
    - But don't be a wimp



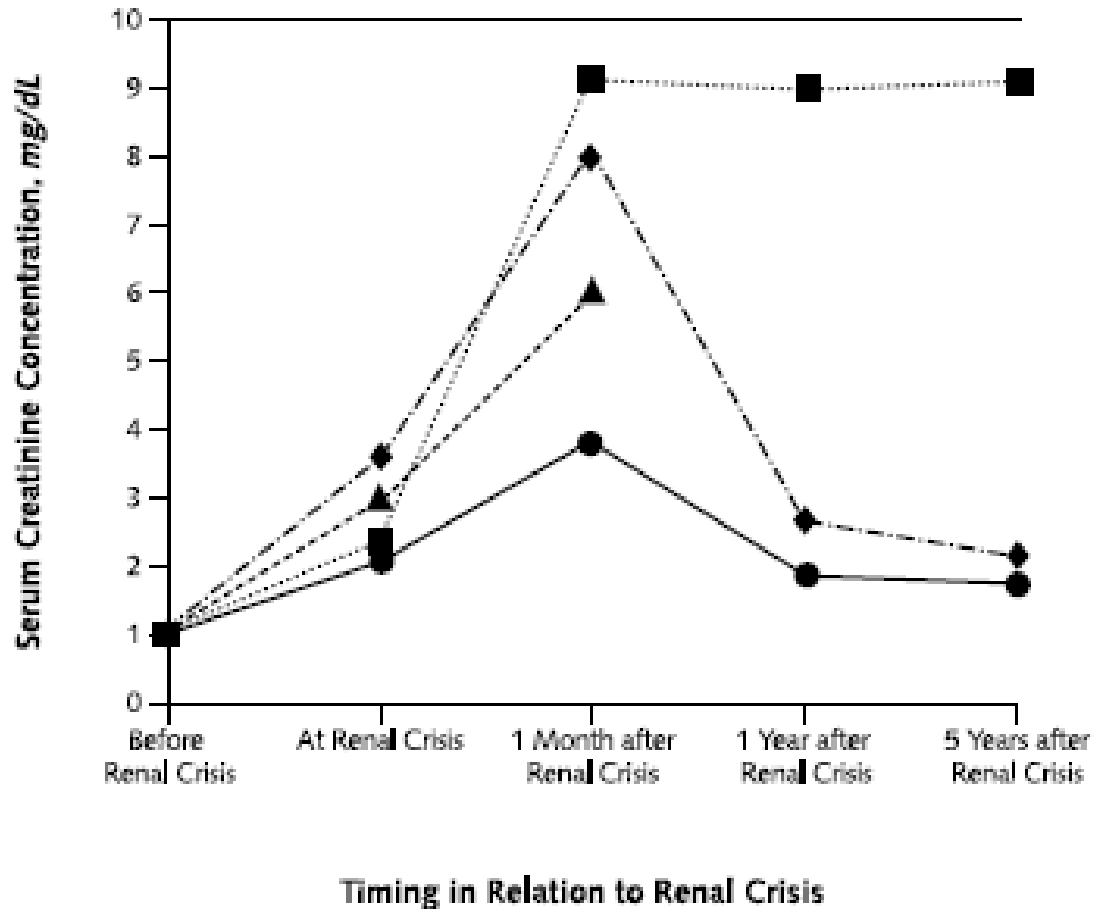
# Treatment of SRC

- Blood pressure control
- Target unclear
  - 20% reduction standard in hypertensive emergencies
- Consider adjuvants to ACEi if not making progress
  - Nitroprusside
  - CCB
  - Vasodilators
  - Avoid Beta-blockers for potential reflex vasospasm

# Treatment of SRC

- Watch
  - Creatinine
  - Potassium
  - Hemoglobin
  - Platelets
  - Other vital organ function
- Consider support with dialysis if necessary
  - Refer early – vascular access can be an issue

# (Renal) Recovery from SRC



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**WHEN TO REFER**

# Nephrologists Perspective

- Rarely unhappy with a consult for a rise in creatinine in SSc patients
  - Double check the creatinine
  - Dip the urine
  - Rehydrate when necessary
  - Check for NSAIDs

# Conclusions

- SRC is bad so...
- Monitor regularly in high risk patients
  - Just diagnosed with diffuse SSc or required prednisone
- Time is kidney so diagnose quickly
  - Refer to your friendly neighborhood nephrologist when there is indecision regarding creatinine
- Jump on the ACEi and aggressively when you're confident in the diagnosis
- Don't lose hope when things aren't going well



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**QUESTIONS?**