

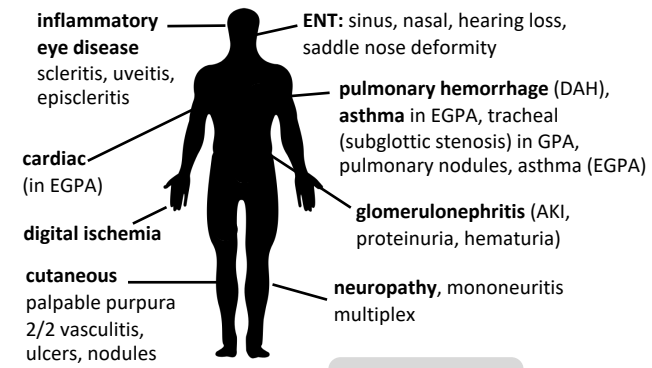
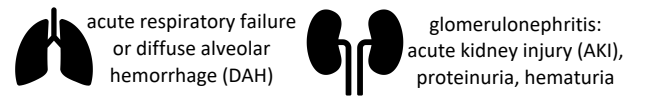
PULMONARY-RENAL SYNDROMES

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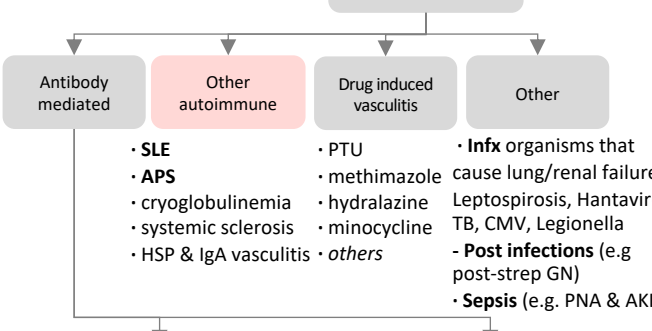

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PRESENTATION:

Pulmonary Renal Syndromes (PRS) are life-threatening diseases with **pulmonary hemorrhage (DAH)** & **renal failure** (glomerulonephritis). Although pulmonary and renal involvement is the defining feature, PRS can affect many organs:



DIFFERENTIAL DIAGNOSIS:



Autoimmune ANCA vasculitis (AAV)

- GPA (granulomatous with polyangiitis)
- EGPA (eosinophilic granulomatosis w polyangiitis)
- MPA (microscopic polyangiitis)

Anti-basement membrane (Goodpasture)

PATHOPHYSIOLOGY:

- AAV**: ANCA **activates primed neutrophils** → vessel endothelial damage & inflammation; Granulomatous inflammation (cell-mediated immunity) in some forms of AAV (GPA, EGPA)
- GBM**: **Auto-antibodies against α3 chain of type IV collagen**, disrupting the basement membrane integrity in lungs and kidney.
- Environmental risk factors: silica exposure (AAV), smoking (GBM)

WORKUP & DIAGNOSIS

- Labs:**
- BMP (quantify renal injury), Coags (r/o coagulopathy)
 - CBC w differential (check eosinophil count for EGPA)
 - Auto-antibodies**: **ANCA antibody**, Anti-GBM antibody
 - Urine: UA, Urine protein to creatinine (UPC) ratio
 - Cardiac: consider BNP, troponin if concern for EGPA
 - ESR and CRP (non-specific, ESR usually low in anti-GBM)

- Other tests:**
- CT chest to evaluate pulmonary involvement
 - Bronchoscopy: confirm DAH, r/o infection
 - Echocardiogram** for EGPA (↓ LVEF, pericardial effusion)
 - PFTs (outpatient; **increased DLCO after recent DAH**)
 - EMG/NCS** for mononeuritis multiplex/neuropathy
- Diagnosis of PRS**: **biopsy** (gold standard) or serologies + symptoms (not-optimal but may be necessary)

AUTO-ANTIBODIES:

- MPA** → usually p-ANCA
- GPA** → usually c-ANCA
- EGPA** → ANCA in 40-50%
- Goodpasture's** → anti-GBM

	ANCA Associated Vasculitis (AAV)			Anti GBM
	MPA	GPA	EGPA	
P-ANCA (MPO)	65%	15%	45%	20%
C-ANCA (PR3)	15%	85%	5%	10%
X-ANCA (elastase, cathepsin, lysozyme, others)	Perinuclear pattern (but not MPO) Seen with medications : Levamisole/cocaine, hydralazine ANCA also seen in many diseases : SLE, RA, PSC, PBC, AIH, IBD (UC > CD)			

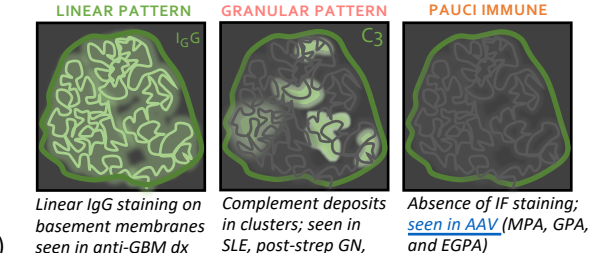


RENAL FINDINGS:

U/A: microscopic or gross hematuria, RBC casts, low grade proteinuria

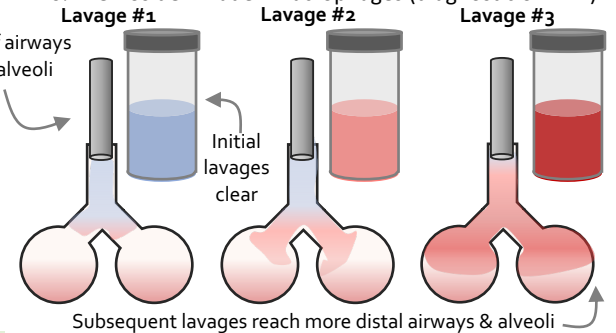
Path: rapidly progressive (crescentic) glomerulonephritis (fibrinoid necrosis, hypercellular glomeruli, & cellular crescents)

IF staining patterns in crescentic GN:



PULMONARY FINDINGS:

- AAV or Goodpasture's** cause **pulmonary capillaritis** leading to **diffuse alveolar hemorrhage (DAH)**
- Diffuse ground glass or consolidative opacities with **sparing or the lung periphery** is typically seen on chest CT.
- BAL** reveals **increasing blood return** in serial lavages and >20% hemosiderin laden macrophages (diagnostic of DAH)



- Subsequent lavages reach more distal airways & alveoli
- Surgical lung biopsy** (not always required) may reveal:
 - linear IgG staining along BM (**anti-GBM**)
 - granular immune complex deposition (SLE & rheum dx)
 - pauci immune (no Ig, immune complex) with neutrophil infiltration of vessels (**GPA**) or eosinophils (**EGPA**).

OTHER FINDINGS:

Skin lesions: palpable purpura, petechiae, ulcerations, & occasionally nodules. Nasal or sinus mucosa often involved in EGPA. Biopsy of skin or nasal mucosa can reveal vasculitis, such as **leukocytoclastic vasculitis of skin**

APPROACH: Initial tx focused on **remission-induction**
Later tx focused on maintenance.

