ANCA Associate Vasculitis

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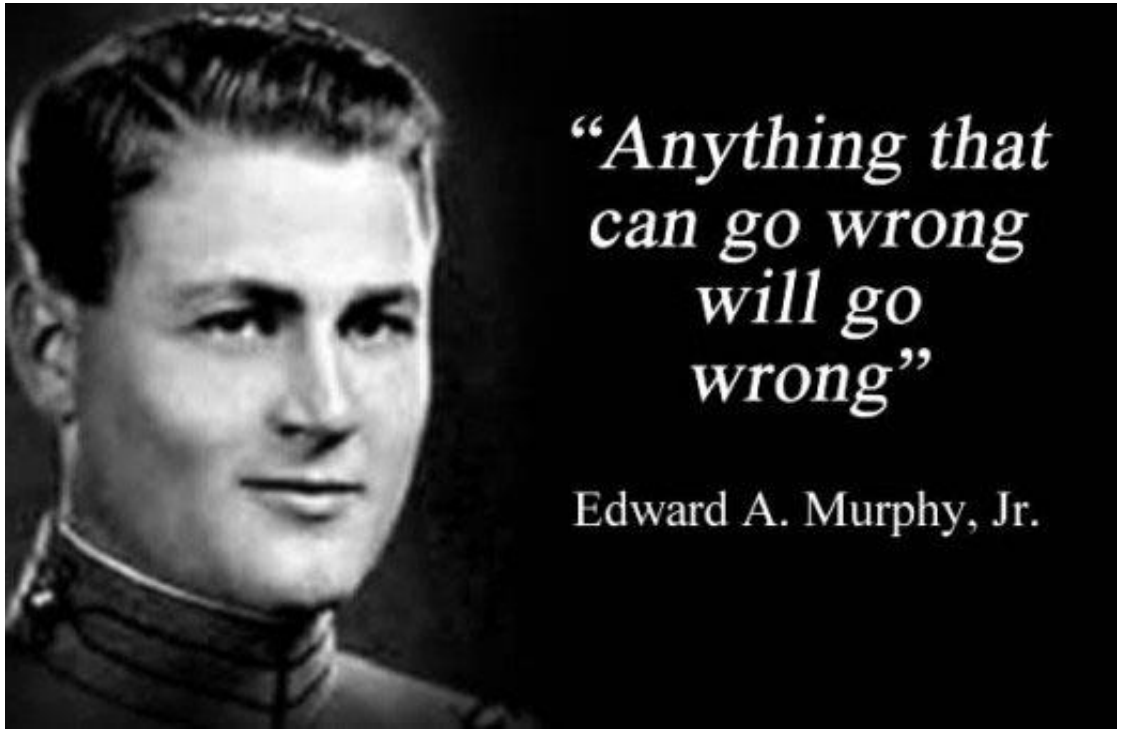
Tehran, 2024

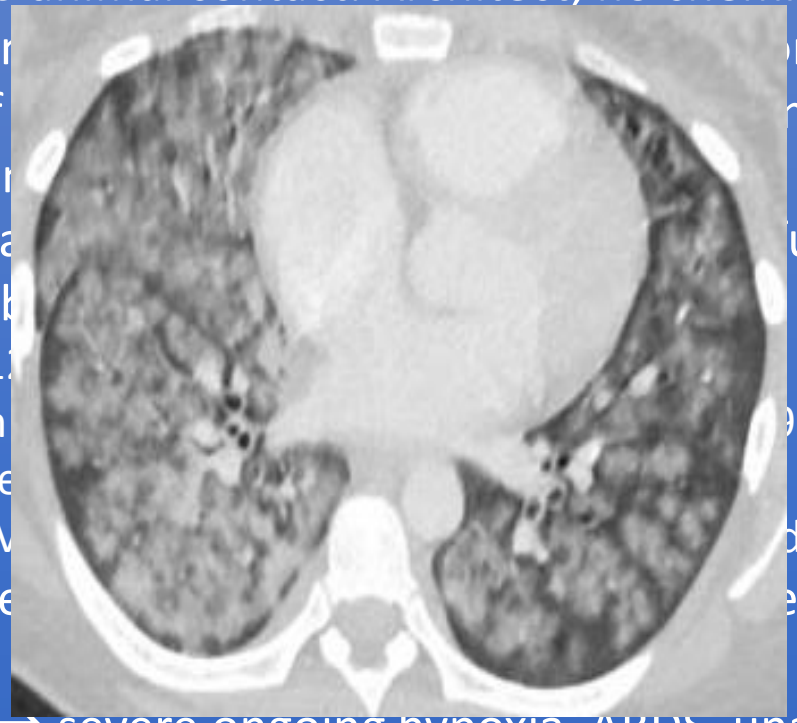

MERCK
Pfizer
Cephalon
gsk
GlaxoSmithKline
Abbott
ALLERGAN
Baxter
GE Healthcare
JANSSEN PHARMACEUTICAL LTD
SOLVAY
Roche
sanofi aventis
SCHWARZ PHARMA
Bristol-Myers Squibb
Wyeth
Zydus Cadila
NOVARTIS
SANDOZ
Alembic
Dabur
Glenmark
None
No AI tools were used to prepare this presentation
INTAS
LYKA
USV
Roche
Project Fusion
Project Fusion

Murphy's Laws

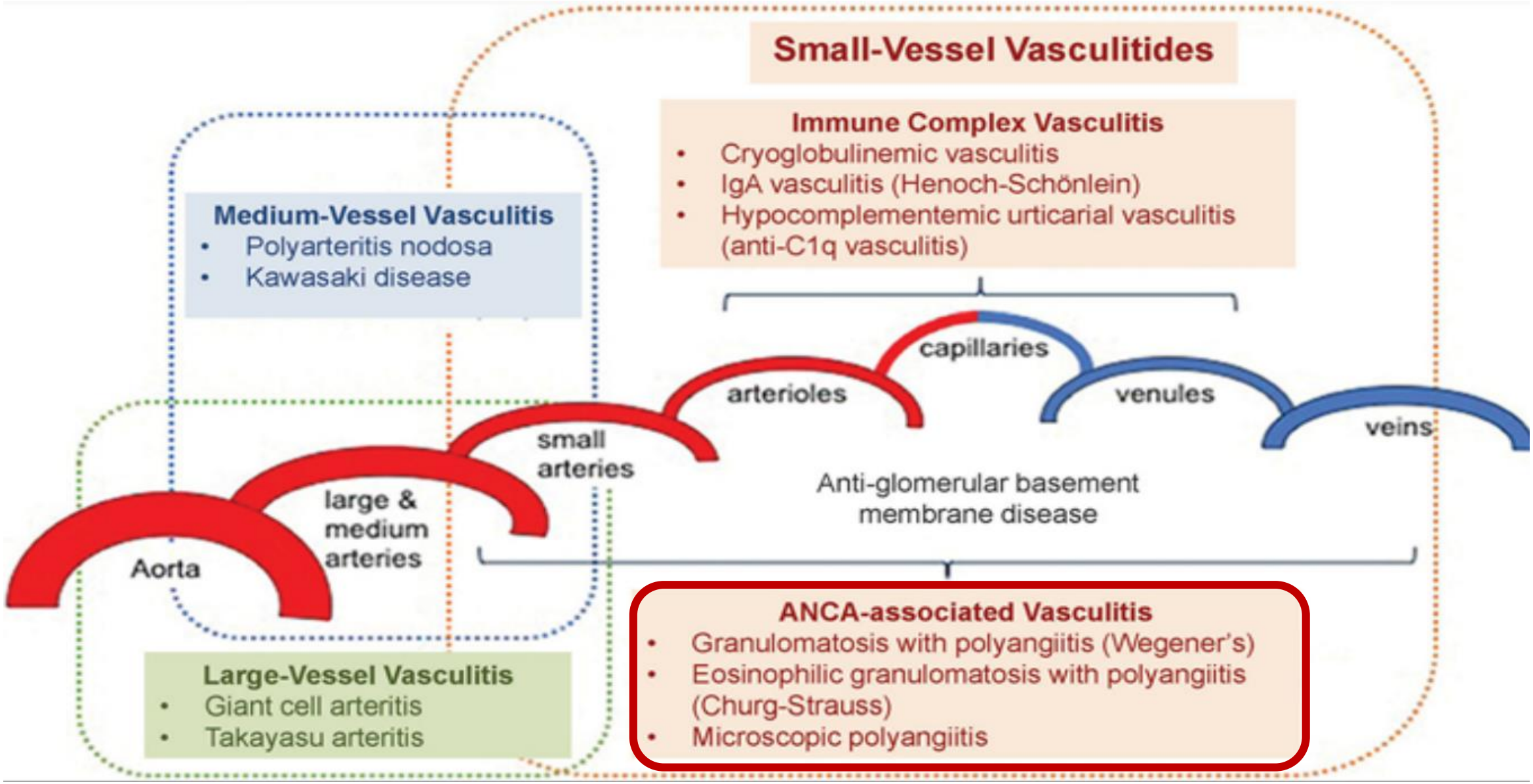
1. In any field of endeavor, anything that can go wrong, will go wrong.
2. Left to themselves, things always go from bad to worse.
3. If there is a possibility of several things going wrong, the one that will go wrong, is the one that will cause the most damage.
4. Nature always sides with the hidden flaw.
5. If everything seems to be going well, you have obviously overlooked something.



- 27 YO single female, nausea, vomiting weakness, fever, and chills
- Recent travel to the north of the country
- Non-smoker, no animal contact. Architect, no chemical contact
- Past h/o AS on r… …matology.
- One episode of … …nent)
- COVID-19 vacci…
- Mother has disa… …use vitiligo.
- Presentation lab…
  - Creatinine 1…
  - Hemoglobin … …90,000
  - + schistocyte…
  - Negative HIV… …d Influenza PCR
- Urgent HD unde… …esis, antimicrobials, and supportive …
- ICU admission → severe ongoing hypoxia, ARDS, unabating DAH
- All available rheumatology w/u were negative
- Increased Ferritin, LDH, CRP, PCT, and IL6
- **?Idiopathic Alveolar Capillaritis**
- Unresponsive DAH and ARDS. aFVII was not agreed upon
- Patient died after 24 days of multidisciplinary care.

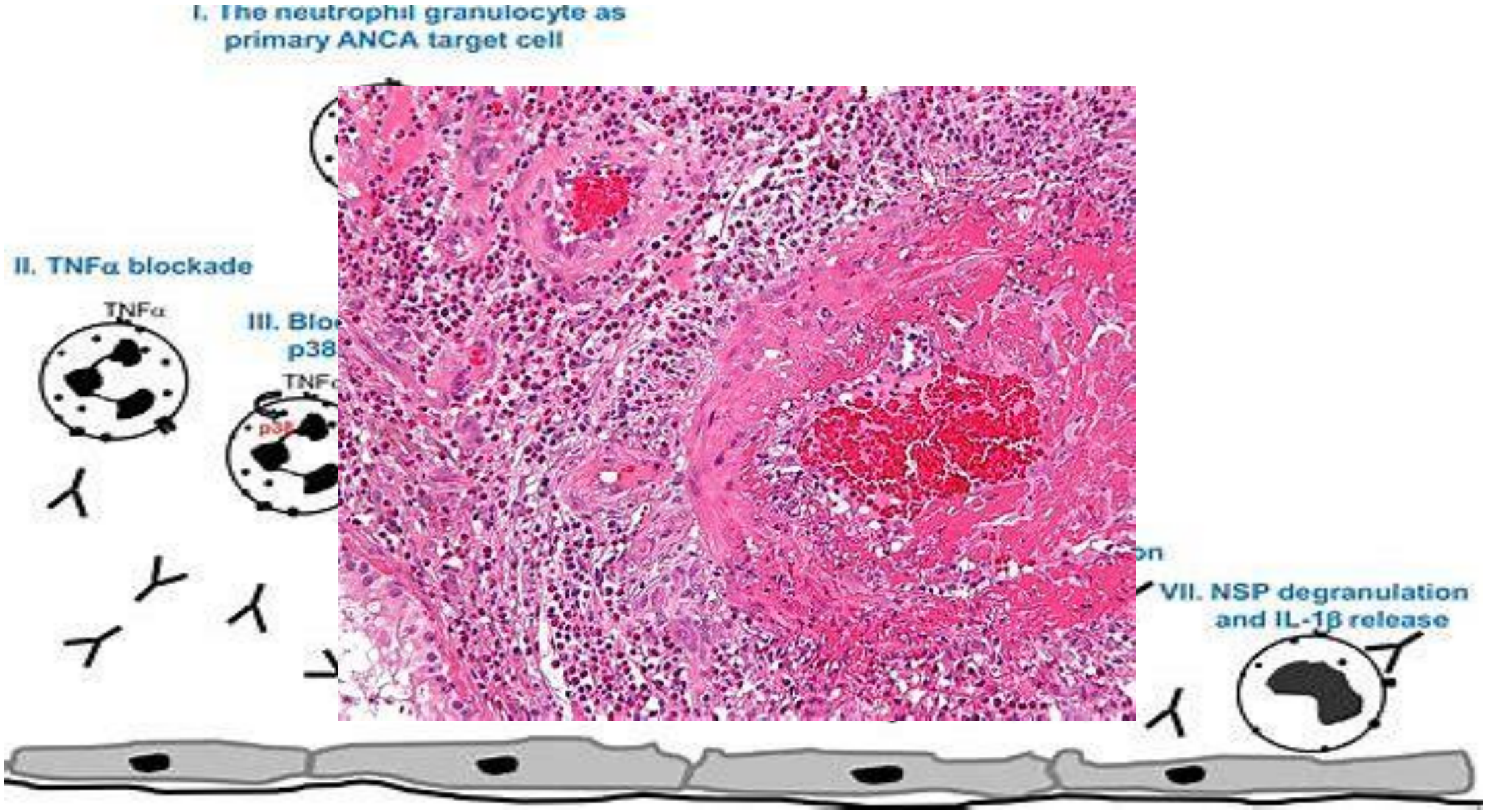


Classification of Vasculitis

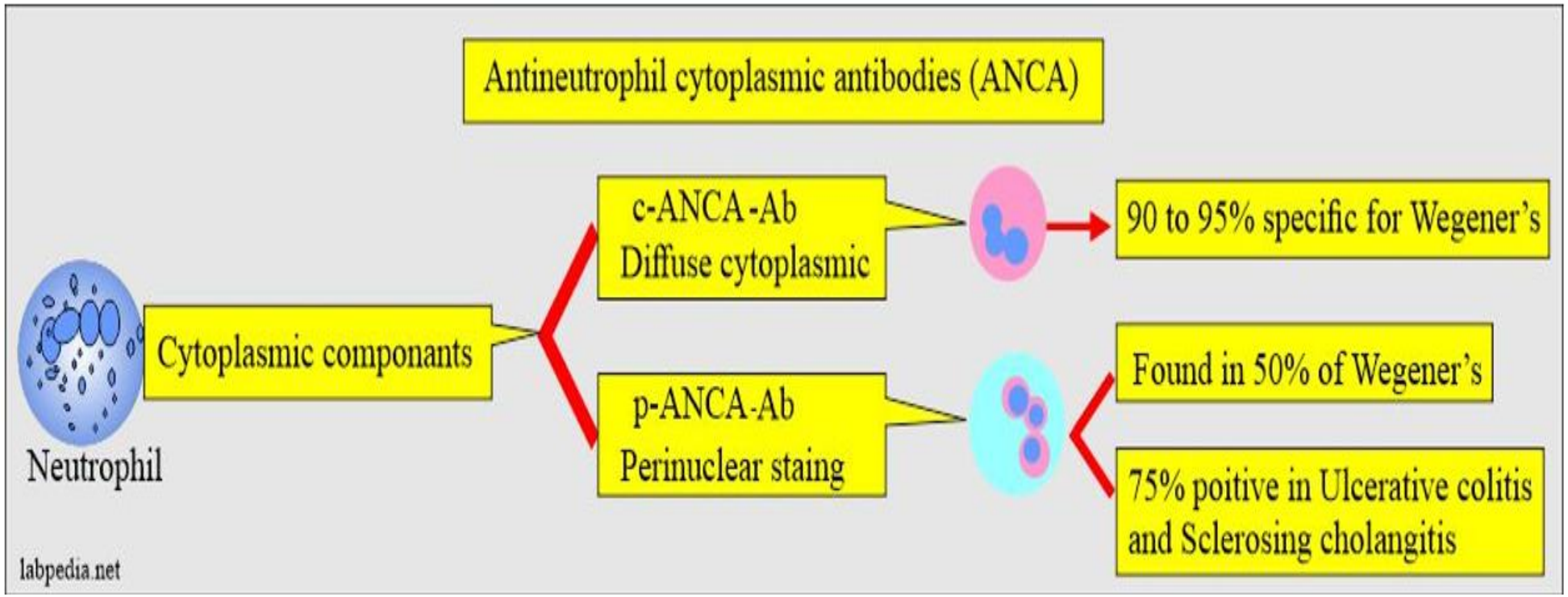


Adapted from Schnabel A, et al. Open Access 2021

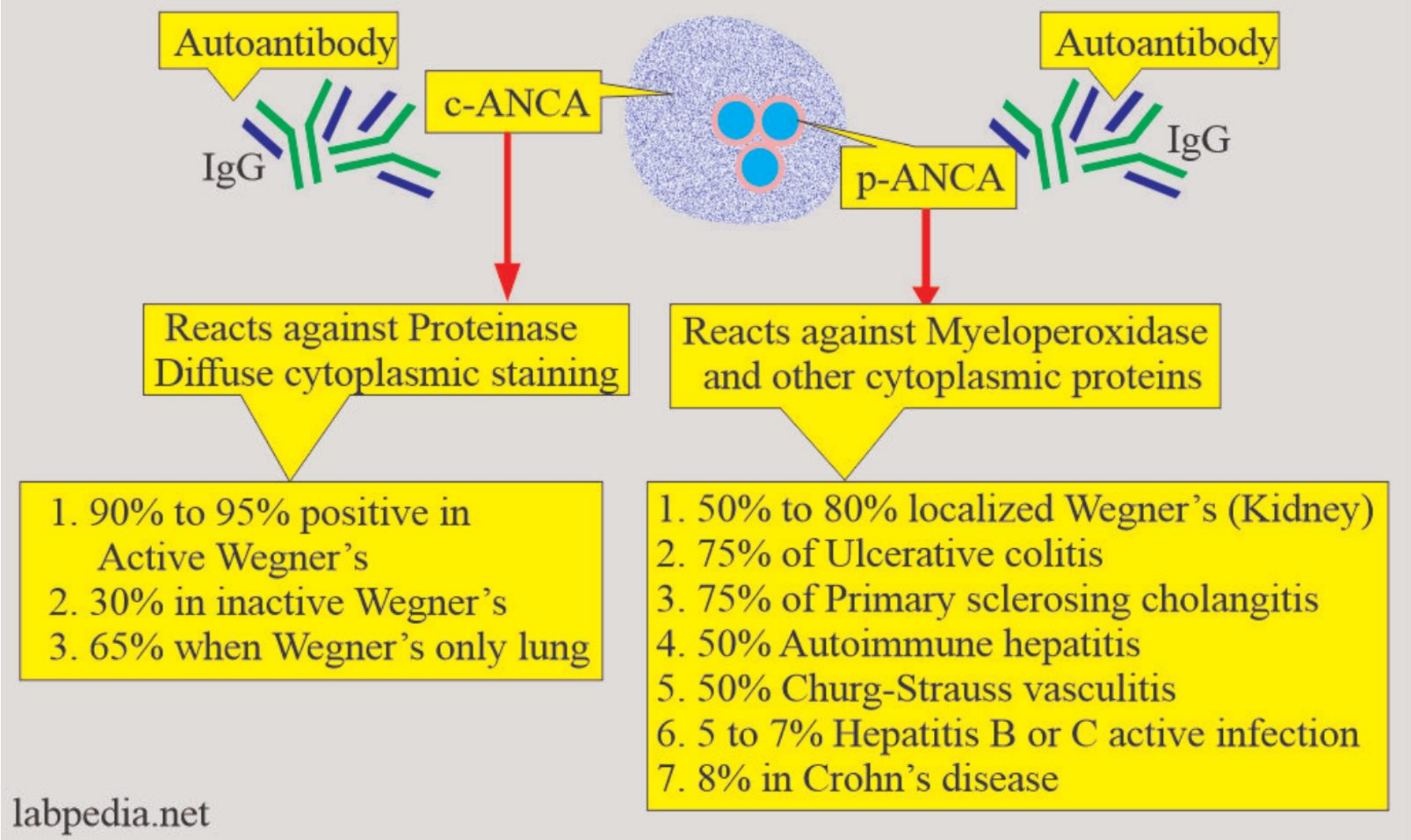
Antineutrophil Cytoplasm Antibody



Antineutrophil Cytoplasm Antibody



ANCA Positivity in Various Diseases



ANCA Specificity in Small Vessel Vasculitis

	ANCA Specificity	
	p-ANCA (MPO)	c-ANCA (PR3)
Granulomatosis with polyangiitis (GPA)	25%	70%
Microscopic polyangiitis (MPA)	60%	35%
Eosinophilic granulomatosis with polyangiitis (EGPA)	30%-40%	Uncommon

EGPA Diagnostic Criteria, Classification, and Definition

Lanham Diagnostic Criteria (1984)[7]

- Asthma
- Eosinophilia >1500/mm^3 or >10% of total WBC
- Evidence of vasculitis involving ≥2 organs

American College of Rheumatology Classification Criteria (1990)[9]

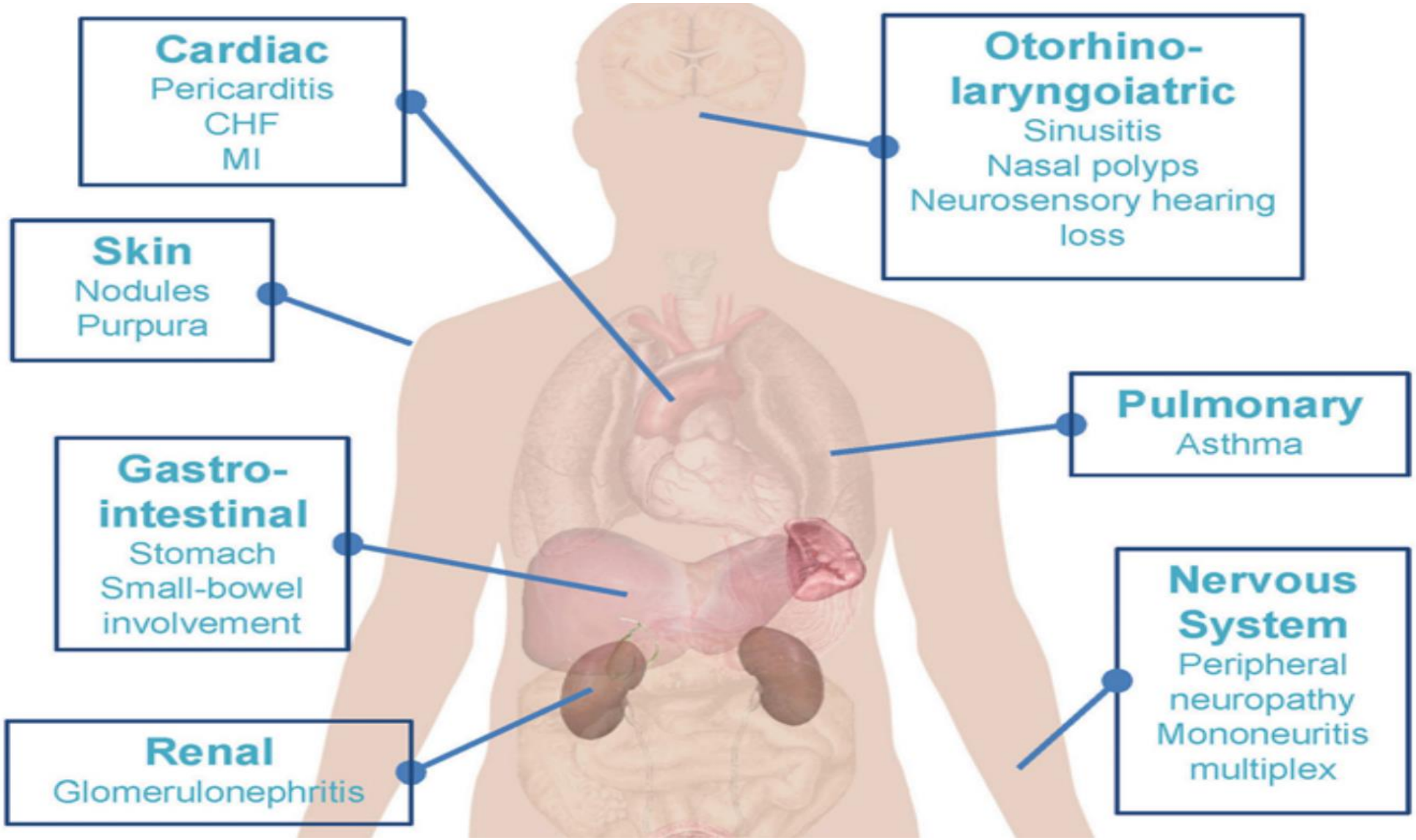
- Asthma
- Eosinophilia (>10% of total WBC)
- Neuropathy
- Pulmonary infiltrates nonfixed
- Paranasal sinus abnormalities
- Extravascular eosinophils

Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides (2012)[3]

- Eosinophil-rich, necrotizing, granulomatous inflammation (often involving respiratory tract)
- Necrotizing vasculitis (small-medium vessel)
- Asthma
- Eosinophilia
- ANCA: more frequent when glomerulonephritis is present

Jennette JC, et al ; Lanham JG, et al ; Masi AT, et al 2013

Potential Manifestations of EGPA



Lanham JG, et al, Baltimore 1984

AAV Pathophysiological Types

- Granulomatosis with polyangiitis (GPA; formerly known as Wegener granulomatosis)
- Eosinophilic granulomatosis with polyangiitis (EGPA; previously known as Churg-Strauss syndrome)
- Microscopic polyangiitis (MPA)
- Other ANCA-associated diseases
  - Drug-induced vasculitis
  - Renal-limited vasculitis

AAV Pulmonary Diseases

Necrotizing granulomatous inflammation

Tracheobronchial inflammation

Pulmonary Capillaritis

Interstitial Lung Disease

Asthma

?

- **Local treatments for subglottic stenosis**
- **The uncertain efficacy of plasma exchanges for alveolar hemorrhage**
- **The potential role of antifibrotic agents in ILD associated with MPA**
- **The use of novel anti-IL5 strategies in EGPA.**

AAV Pulmonary Diseases

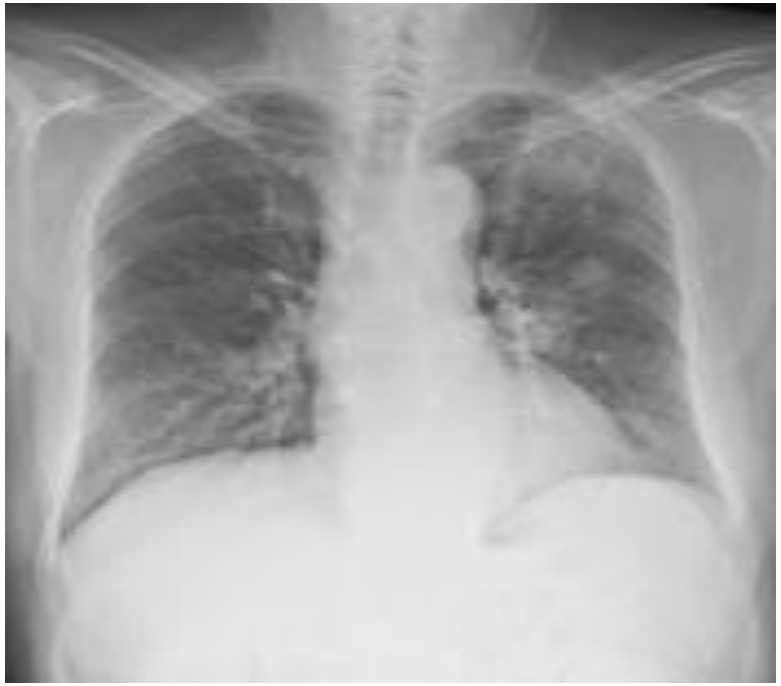
Three main clinicopathologic syndromes

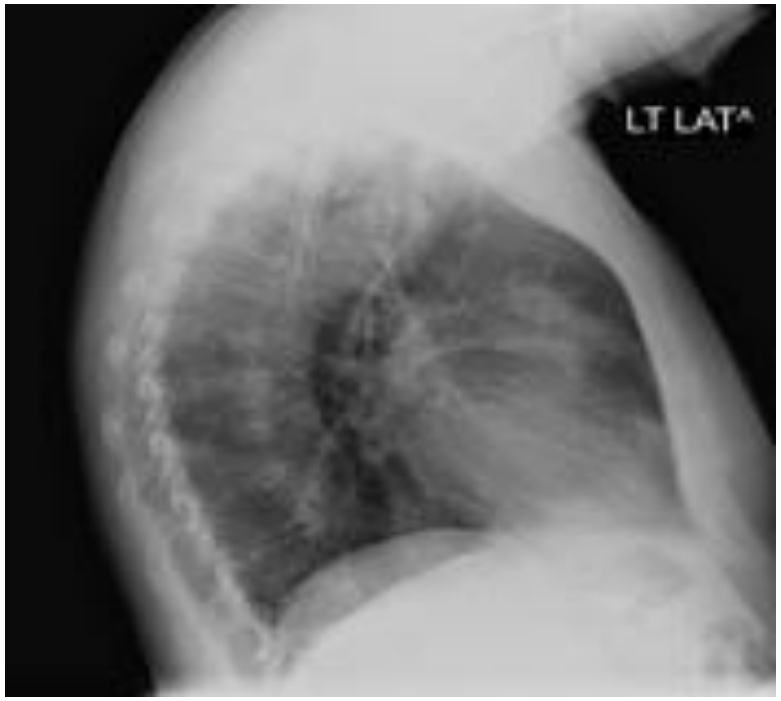
- Granulomatosis with polyangiitis (GPA 67-85%)
- Microscopic polyangiitis (MPA up to 60%)
- Eosinophilic granulomatosis with polyangiitis (EGPA 95% + Asthma)

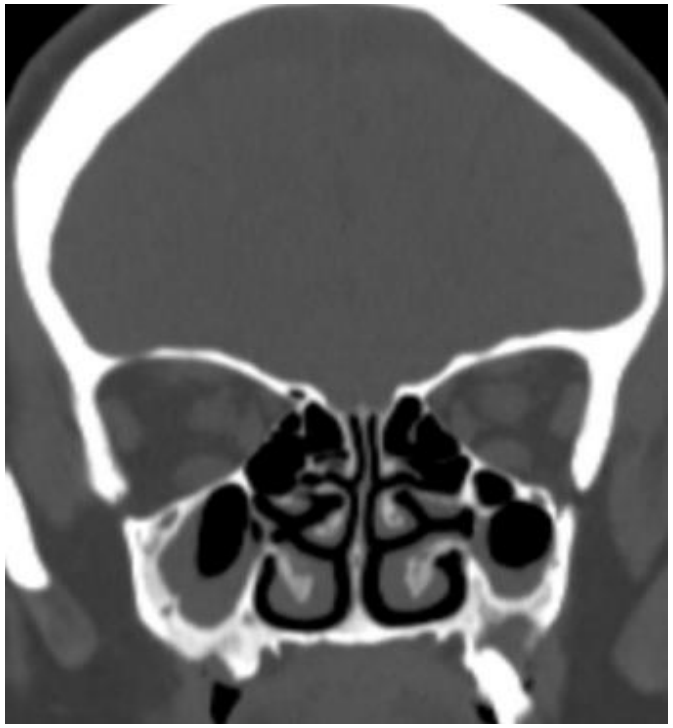
Up to 61% ICU mortality

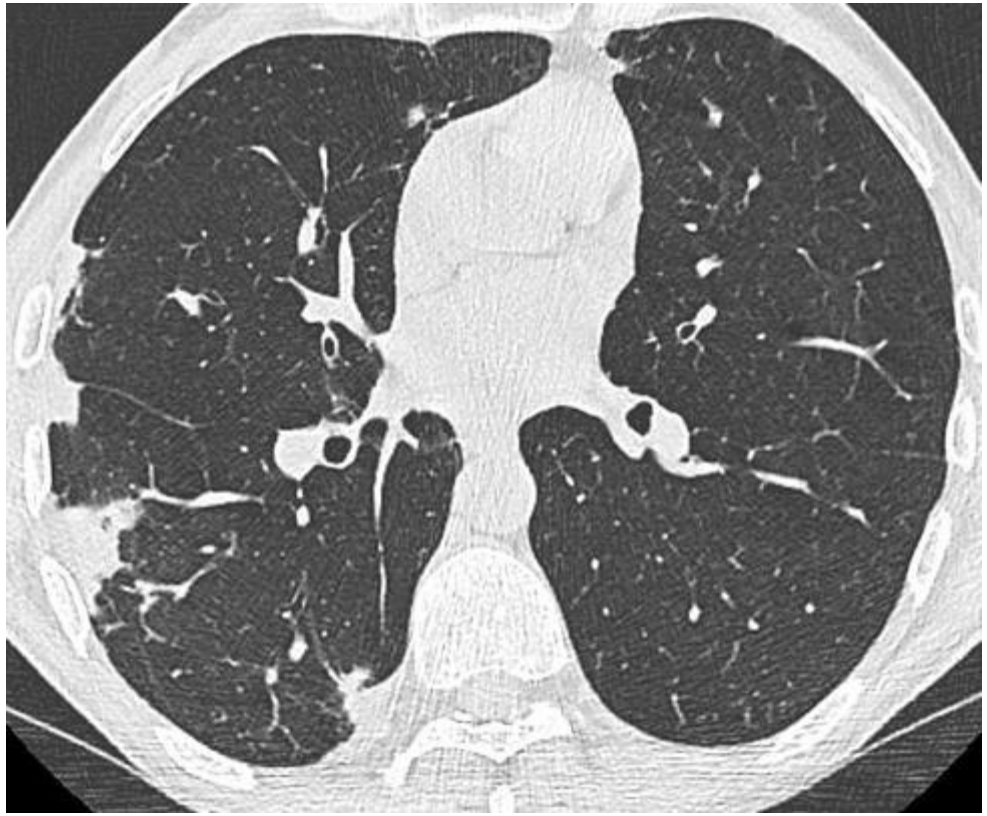
AAV Pulmonary Diseases

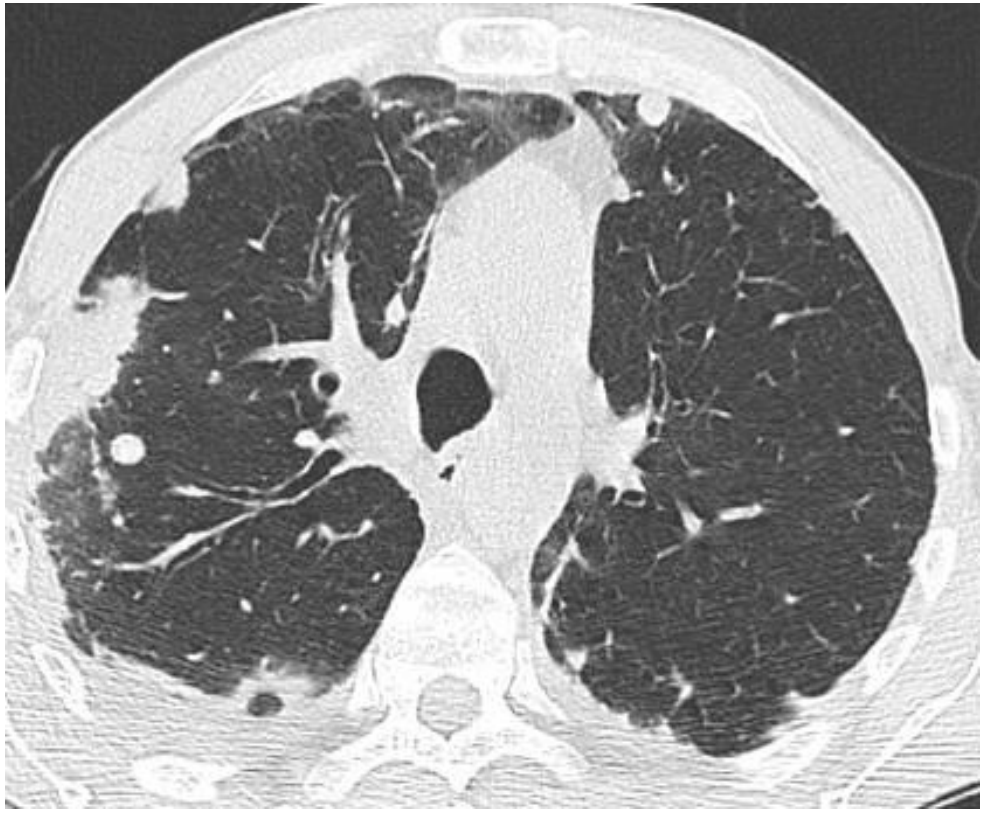
	GPA	MPA	EGPA
Nodules	50% (a)	7% (b)	14% (b)
Tracheobronchial involvement	15-55% (c)	50% (h)	Rare
DAH	22-30% (d)	25% (i)	4% (e)
ILD	23% (f)	45% (f)	39% (e)
Asthma	8% (b)	5% (b)	95-100% (g)

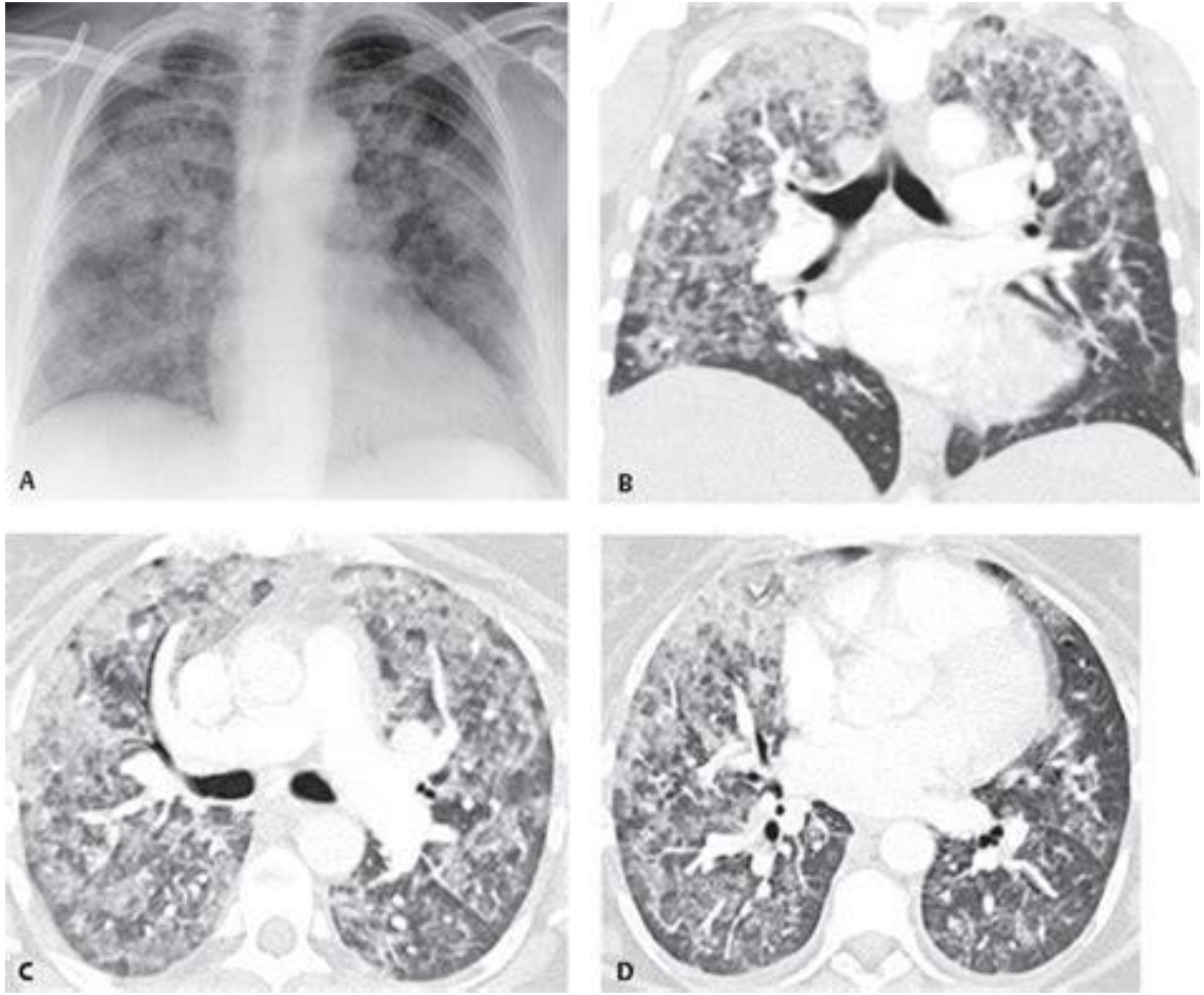



LT LAT^

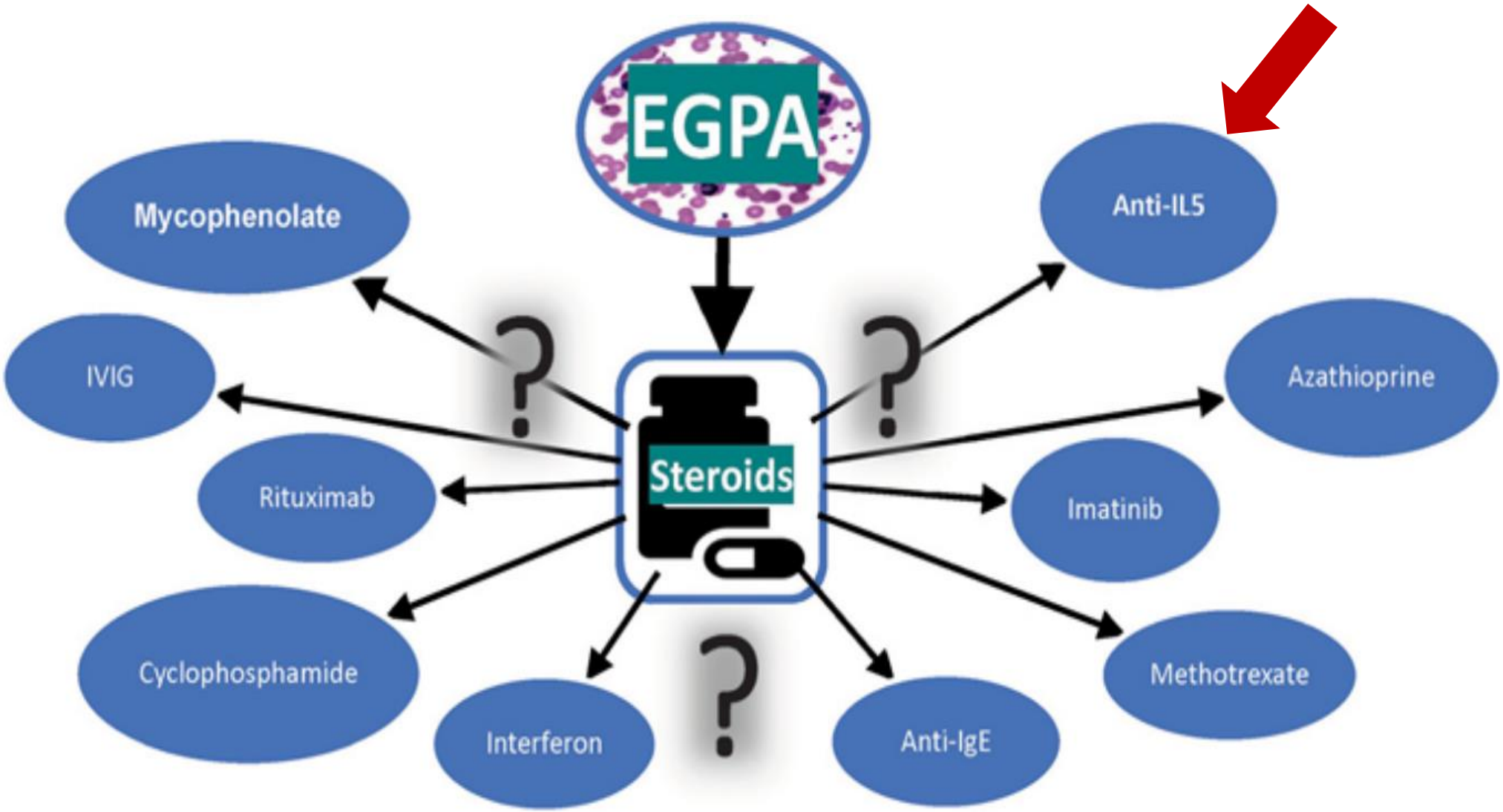







A
B
C
D

Potential Steroid Sparing Management



AAV Pulmonary Diseases

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