



# RhAPP

RHEUMATOLOGY ADVANCED  
PRACTICE PROVIDERS

## Inaugural National Conference

**December 3 – 5, 2020**

VIRTUAL CONFERENCE



**RhAPP**

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# Faculty Disclosures

## **Tiffany Clark, RN, MSN, CNP**

- No relationships to disclose

## **Naomi Amudala, MSN**

- No relationships to disclose



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# Granulomatosis with polyangiitis: Physical Exam

Tiffany Clark RN, MSN, CNP

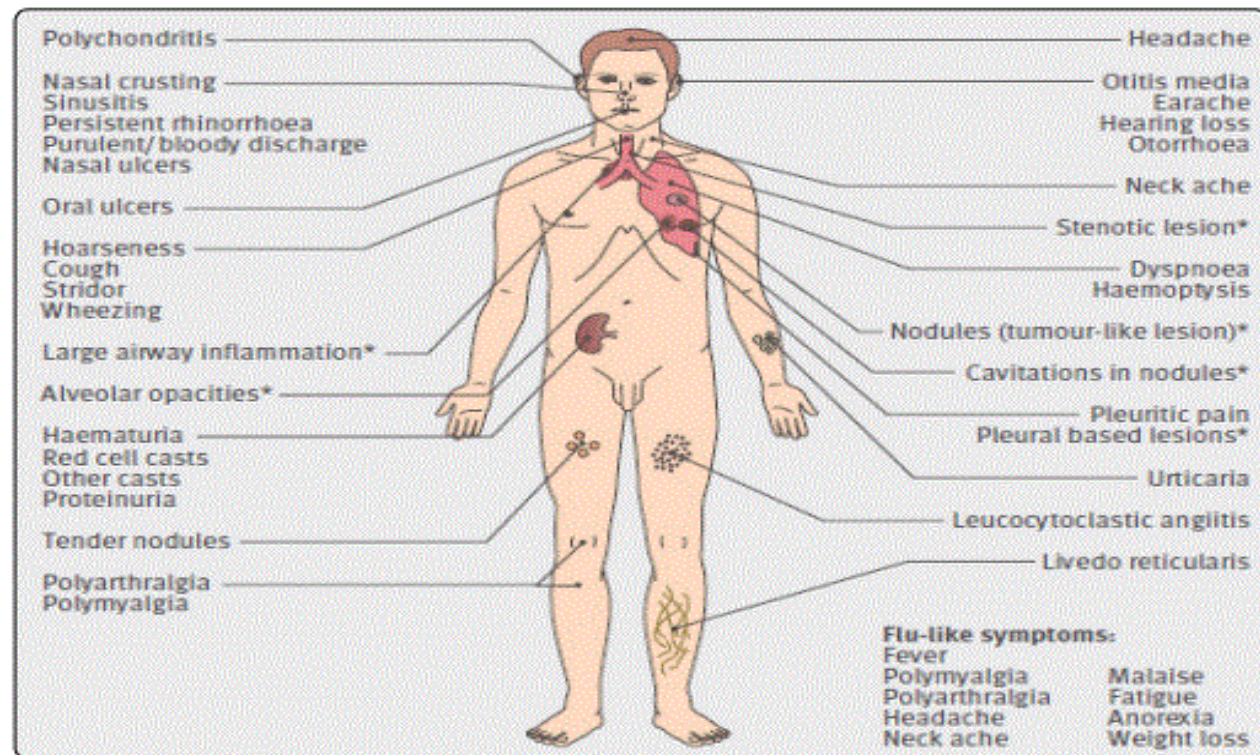
Cleveland Clinic

Orthopedic and Rheumatology Institute

# Physical Exam for suspected GPA

What is needed->

- Patient gown!
- Ear and nasal speculum
- Stethoscope
- Tuning fork



**Figure 2.** Clinical features of ANCA associated vasculitis [3].

# GPA: Nasal Exam

- Critical element
- Nasal speculum required
- Tilt head back, gently exam inner mucosa of nares
- Nasal: crusting, blood (epistaxis), “cobblestone appearance/granular/friable”, nasal septal perforation (trans lumination of light), saddle nose appearance (late, ? mimic) , nasal ulcers, nasal chondritis
- Oral: ulcerative stomatitis, hyperplastic gingivitis, ulcerations



# GPA- Nasal Exam

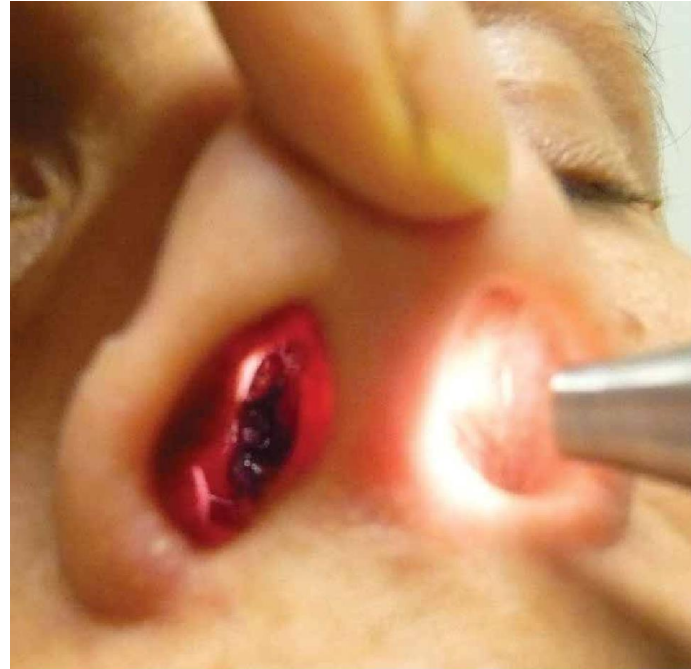


Figure 3. Nasal septum perforation in a patient with polyangiitis (GPA).

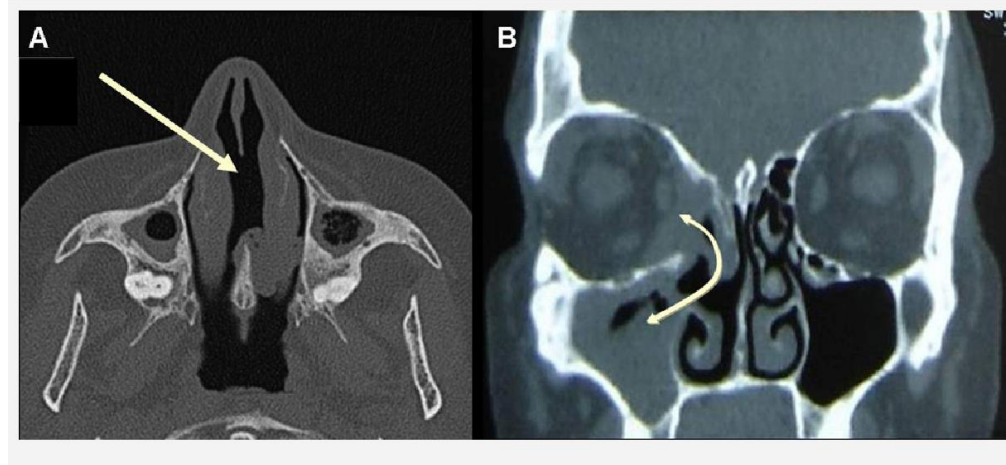
[https://en.wikipedia.org/wiki/Granulomatosis\\_with\\_polyangiitis](https://en.wikipedia.org/wiki/Granulomatosis_with_polyangiitis)

<https://www.semanticscholar.org/paper/Updates-in-ANCA-associated-vasculitis.-Pagnoux/c47def732ecb8a99a303d5eaaff6f6a5f23fee73/figure/1>

# GPA: Sinus exam

## Sinus

- Thick fluid filled sinus cavities
- Recurrent “sinus” infections
- Dullness to percussion of sinuses



# GPA : Orbital Exam

- Conjunctivitis, Scleritis, Episcleritis
- Nasal-Lacrimal gland obstruction- ? Tearing
- Orbital pseudo tumor- check ocular motions, ?  
Double vision (diplopia), loss of vision, color changes, \* proptosis of eye
  - Strangulation of optic nerve
  - Entrapment of extra ocular muscles

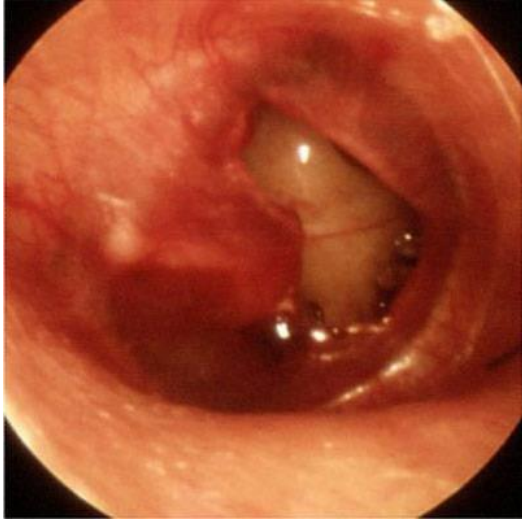
# GPA: Orbital



# GPA: Ear

- Must use ear speculum
- “recurrent ear infections”
- Visual inside ear- Fluid filled (serous or suppurative otitis media), red, draining, TM perforation, ? chondritis of ear
- Inner ear changes: hearing loss (sensorineural deafness- hearing test required), vertigo

# GPA: Ear



# GPA: Pulmonary Exam

- Stethoscope
- Wheezing, crackles, diminished lung sounds
  - Infiltrates, nodules, hemorrhage
- Stridor over the tracheal air way (SG stenosis)
- Percussion: dullness to percussion?
- Cough, SOB, **hemoptysis**



# GPA: Skin

- Be a detective
- Leukocytoclastic vasculitis, splinter hemorrhages, nodules (EN), Livedo reticularis, urticarial rashes, gangrene, palpable purpura





# GPA: Joints- Muscles

- Second most reported symptoms
- Inflamed swollen joints, may be migratory.
- Weakness- strength exam – denote upper and lower muscle groups, ? Myositis
- May mimic RA features- has been reported in up to 2/3'rd of patients
- 90% of patients with Rheumatologic symptoms have a generalized form of the disease.

# GPA: Neurological Exam

- Mononeuritis multiplex
- Sensory and motor exam upper and lower.
  - Foot drop
- Tuning fork vibrations
- GPA urgent issues, once nerves are inflamed and damage – may be permanent. Significant source of chronic damage
- CNS :mass-like lesions, rare- diabetes insipidus, meningitis

# GPA: Cardiac

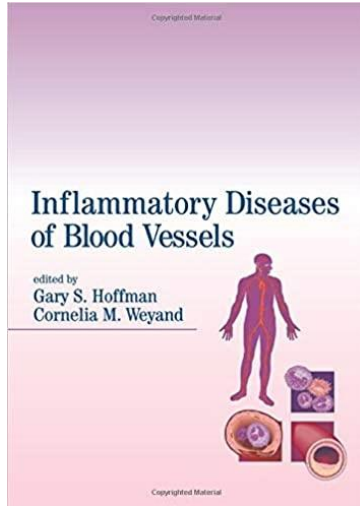
- Myocarditis, Pericarditis, Endocarditis
- Heart sounds, ? Distant

# Know your urgent findings

- GPA can progress rapidly
- GPA may lead to urgent life threatening manifestations quickly
- GPA can be associated with severe chronic irreversible damage and severely impact quality of life

# References

- *Inflammatory Disease of Blood vessels* (G. Hoffman, C. Weyand). 2002 Marcel Dekker, Inc.





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# Granulomatosis with polyangiitis: Laboratory and Diagnostic work-up

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# Initial work-up for suspected GPA

## Laboratories

- CBC w/ differential
- Comprehensive metabolic panel
- Urinalysis
- ESR
- CRP
- ANCA

# Initial work-up for suspected GPA

## Laboratories

- CBC w/ differential
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Anemia  
Thrombocytosis  
Leukocytosis



# Initial work-up for suspected GPA

## Laboratories

- CBC w/ differential
- Comprehensive metabolic panel
- Urinalysis
- ESR
- CRP
- ANCA



Elevated creatinine

# Initial work-up for suspected GPA

## Laboratories


- CBC w/ differential
- Comprehensive metabolic panel
- Urinalysis
- ESR
- CRP
- ANCA



Hematuria  
Red blood cell casts

# Initial work-up for suspected GPA

## Laboratories

- CBC w/ differential
  - Comprehensive metabolic panel
  - Urinalysis
  - ESR
  - CRP
  - ANCA
- 

Non-specific markers of inflammation

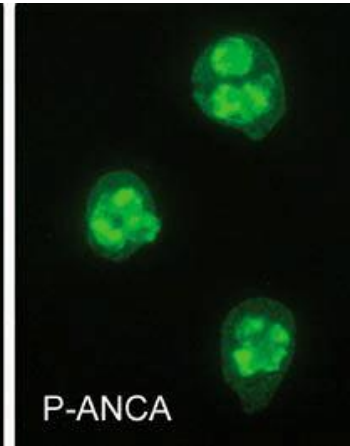
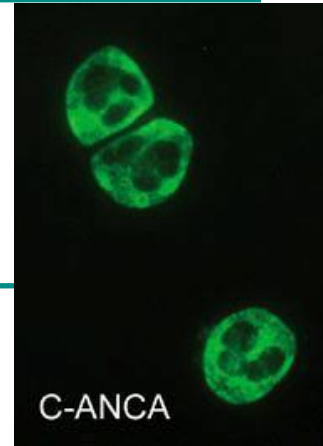
# ANCA interpretation

## Perinuclear staining pattern (P-ANCA)

Myeloperoxidase (MPO) antigen

## Cytoplasmic staining pattern (C-ANCA)

Proteinase 3 (PR3) antigen



<https://www.vasculitis.org.uk/about-vasculitis/what-is-anca>

## Atypical ANCA

- Stains against other antigens
- Dual positive PR3 and MPO

# ANCA interpretation

10% of patients are ANCA negative

75-80% of patient with renal limited vasculitis are MPO positive

PR3 positivity is associated with higher risk of relapse

# Initial work-up for suspected GPA



Chest x-ray



Chest CT scan



Neck CT scan



Head CT scan

[https://www.google.com/url?sa=i&url=https%3A%2F%2Fen.wikipedia.org%2Fwiki%2FChest\\_radiograph&psig=AOvVaw2Q4VDZLzQ\\_0Yp\\_iDLpaW\\_7Z&ust=1606063144187000&source=images&cd=vfe&ved=2ahUKEwjrmrevKiZTtAhUnVd8KHe1HC8sQr4kDegUIARDMAQ](https://www.google.com/url?sa=i&url=https%3A%2F%2Fen.wikipedia.org%2Fwiki%2FChest_radiograph&psig=AOvVaw2Q4VDZLzQ_0Yp_iDLpaW_7Z&ust=1606063144187000&source=images&cd=vfe&ved=2ahUKEwjrmrevKiZTtAhUnVd8KHe1HC8sQr4kDegUIARDMAQ)  
<https://www.sciencephoto.com/media/728474/view/normal-sinuses-ct-scan>  
<https://radiopaedia.org/cases/normal-chest-ct-lung-window-1>

# Initial work-up for suspected GPA

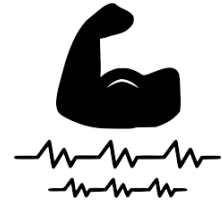
## Other Diagnostics



**Audiogram**



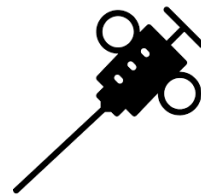
**Laryngoscopy**



**EMG**

# Initial work-up for suspected GPA

## Other Diagnostics - Biopsy



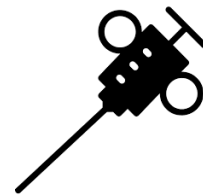
### Skin

- Try to get lesions between 24-48 hours old
- Usually obtained via punch biopsy
- Findings – leukocytoclastic vasculitis
- Direct immunofluorescence findings should be negative – “pauci-immune”



# Initial work-up for suspected GPA

## Other Diagnostics - Biopsy



### Renal

- Pauci-immune glomerulonephritis
- Majority of these patients will be ANCA positive

### Lung

- Biopsy appropriate if suspicious for other causes (infection or malignancy)
- Not advised if patient has alveolar hemorrhage

### Nasal/Sinus

- Findings usually non-specific

# Summary – Diagnostic work-up

## Labs

- CBC w/ diff, metabolic panel, ANCA, ESR/CRP, UA

## Imaging

- CT head, neck, chest

## Other

- Laryngoscopy, EMG, audiogram

## Biopsy

- Skin least invasive biopsy site
- Other sites: kidney, lung, nasal/sinus (less specific)



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## Thank you

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