UNIVERSITY OF MARYLAND

Relapsing Polychondritis

Clinical Pearls for the Practicing Rheumatologist

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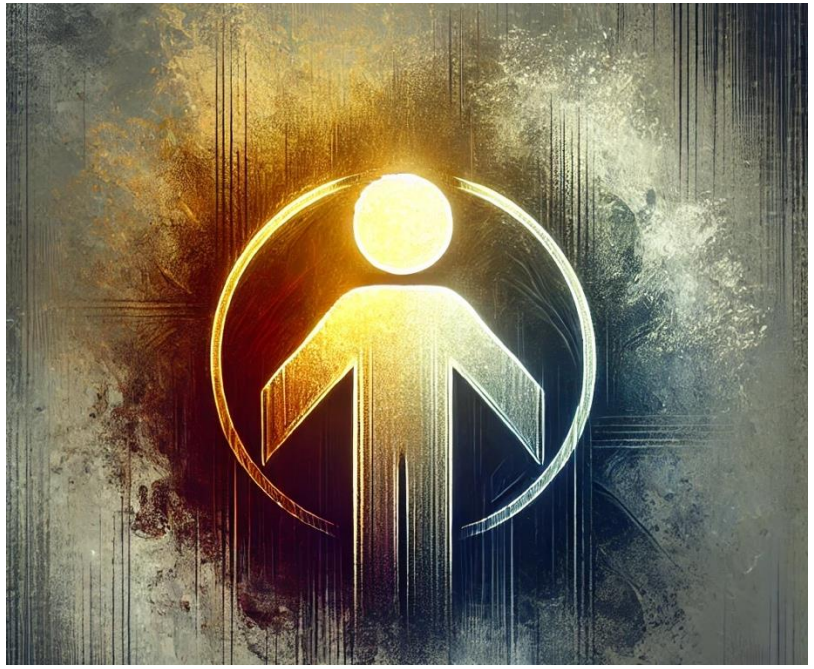
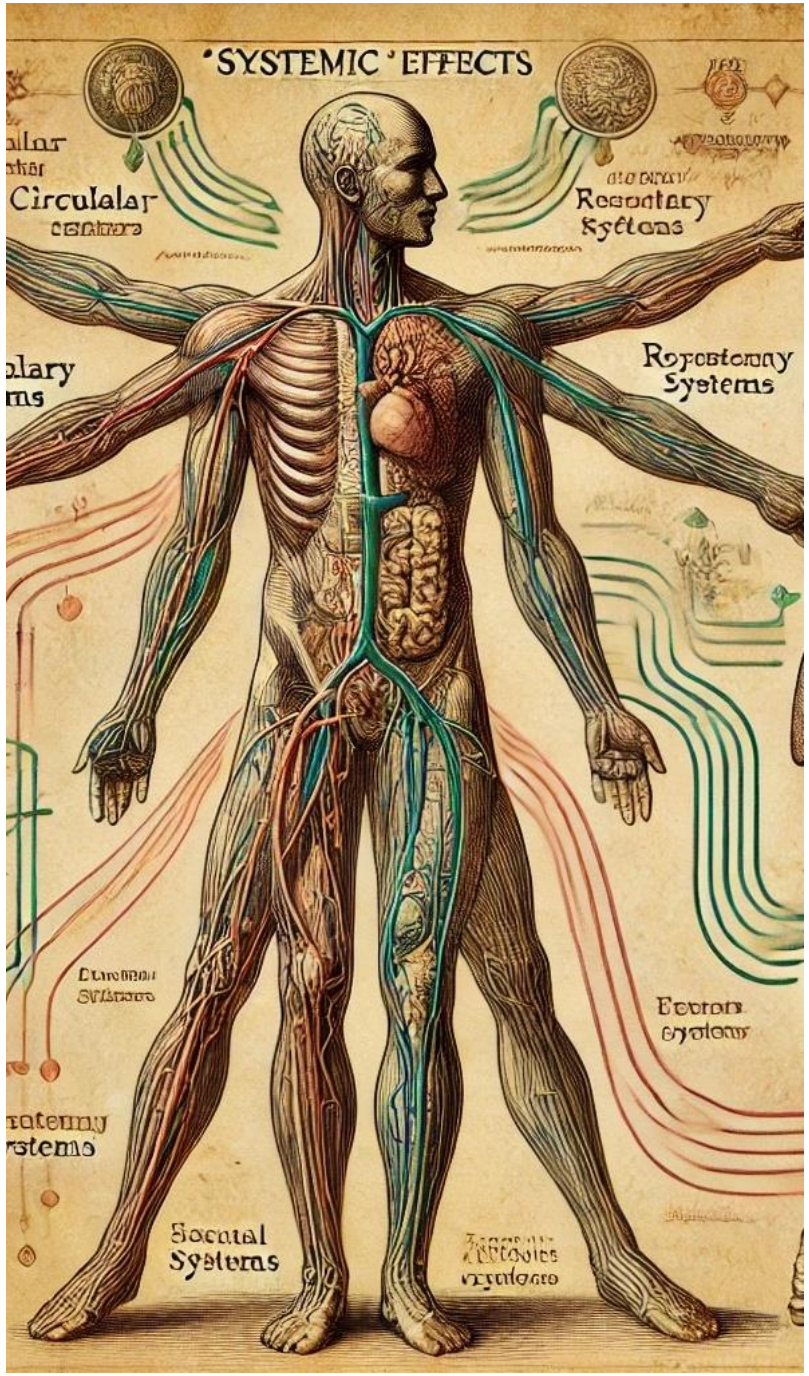
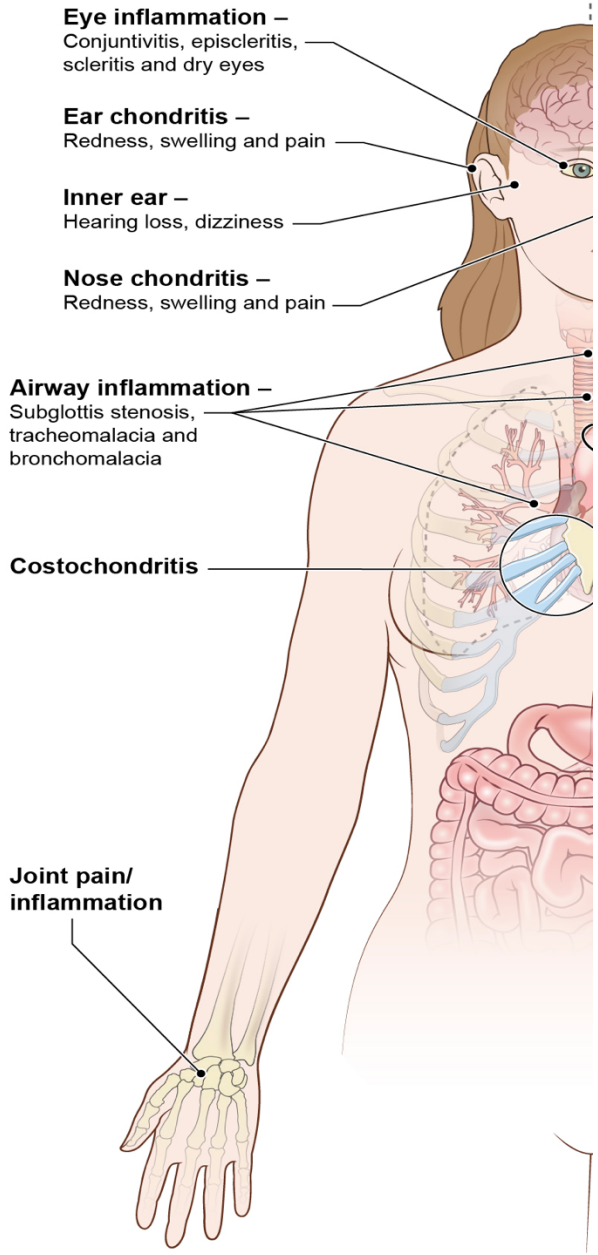
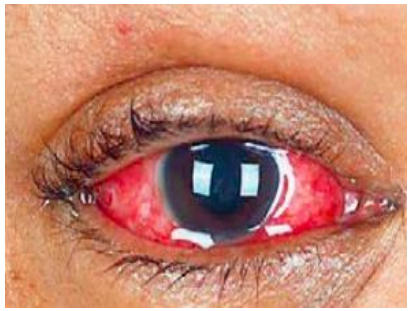
What is Relapsing Polychondritis?

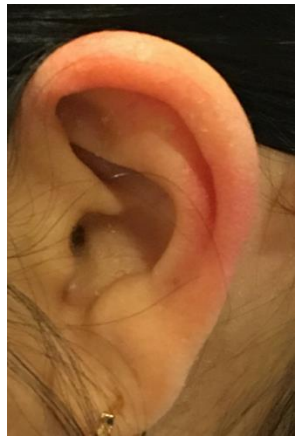


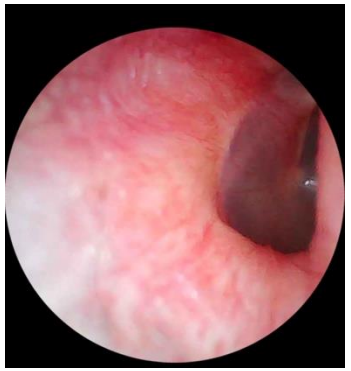

'SYSTEMIC' EFFECTS
Circulalar
Respiratory
Systems
Reproductory
Systems
Social
Systems

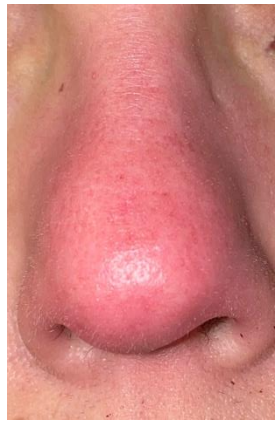
Common manifestations

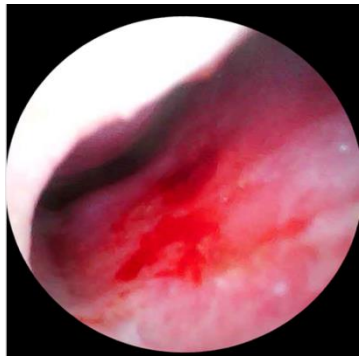


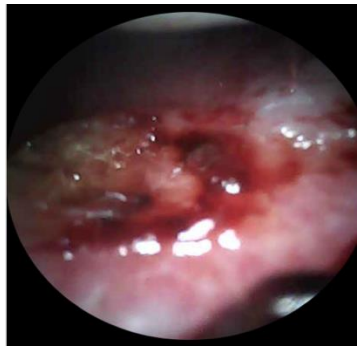


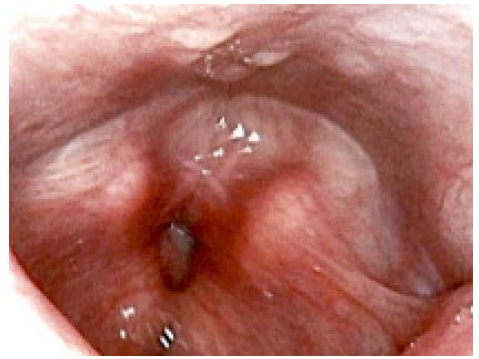


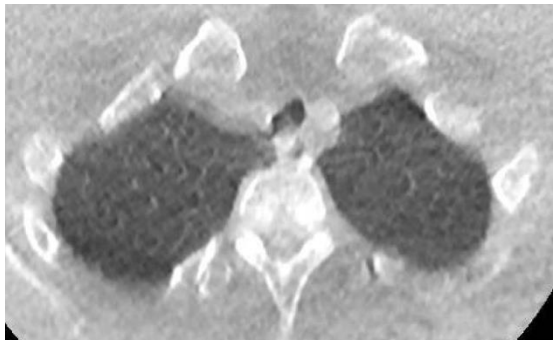


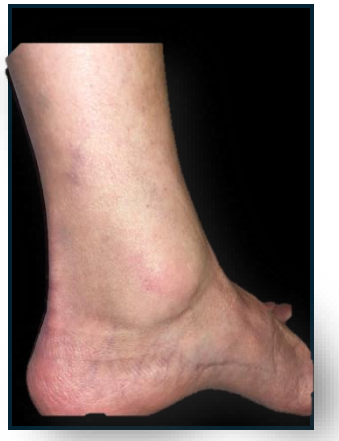








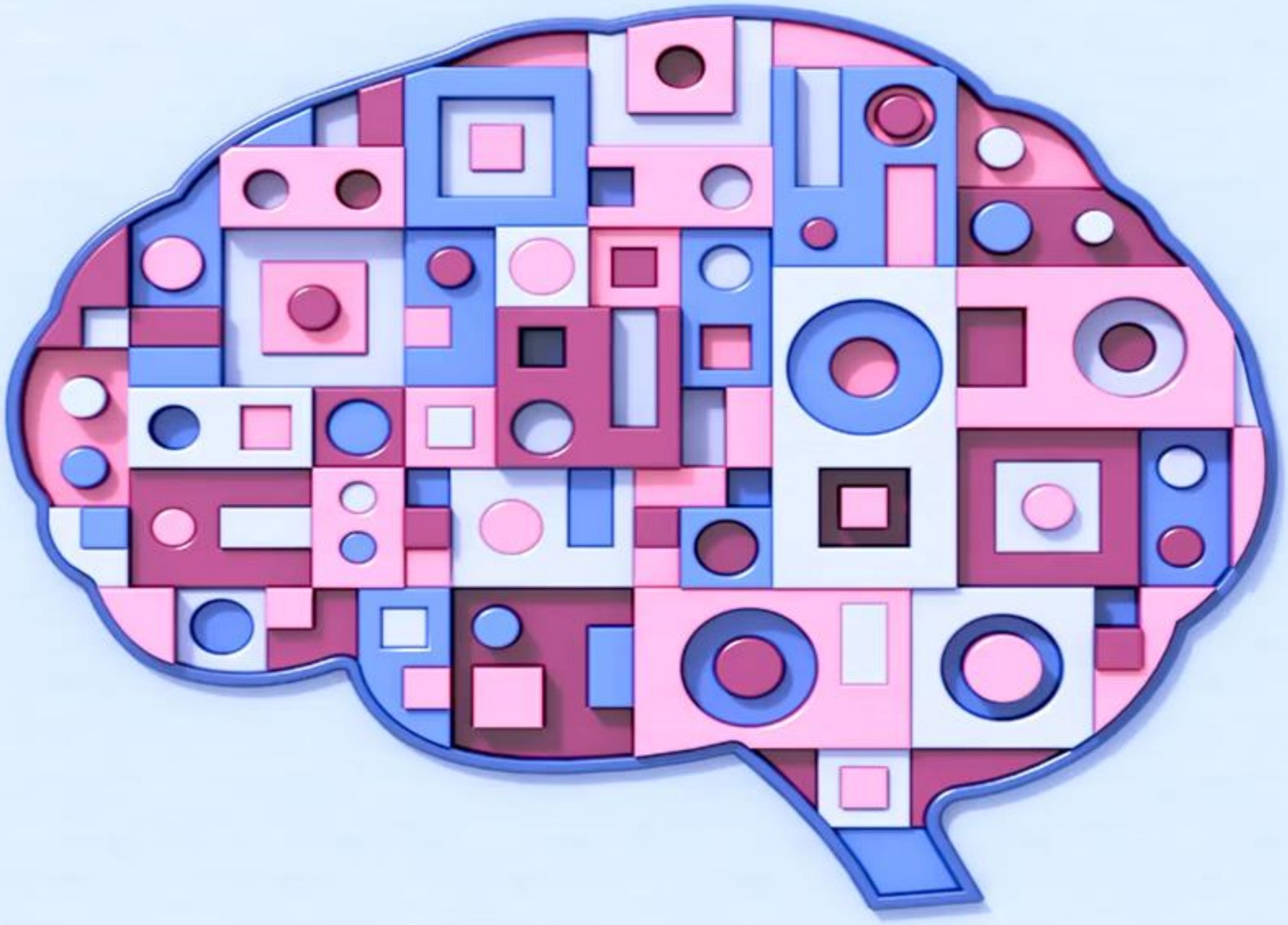




RP Diagnosis is Purely Clinical

Clinical Symptoms (CS)	McAdam's Criteria	Damiani's Criteria
• Bilateral auricular chondritis	3 of 6 symptoms	3CS
• Nasal cartilage inflammation		1CS + biopsy
• Respiratory tract chondritis		2CS+ response to steroids or dapsone
• Non-erosive seronegative polyarthritis		
• Ocular inflammation		
• Audio-vestibular involvement		

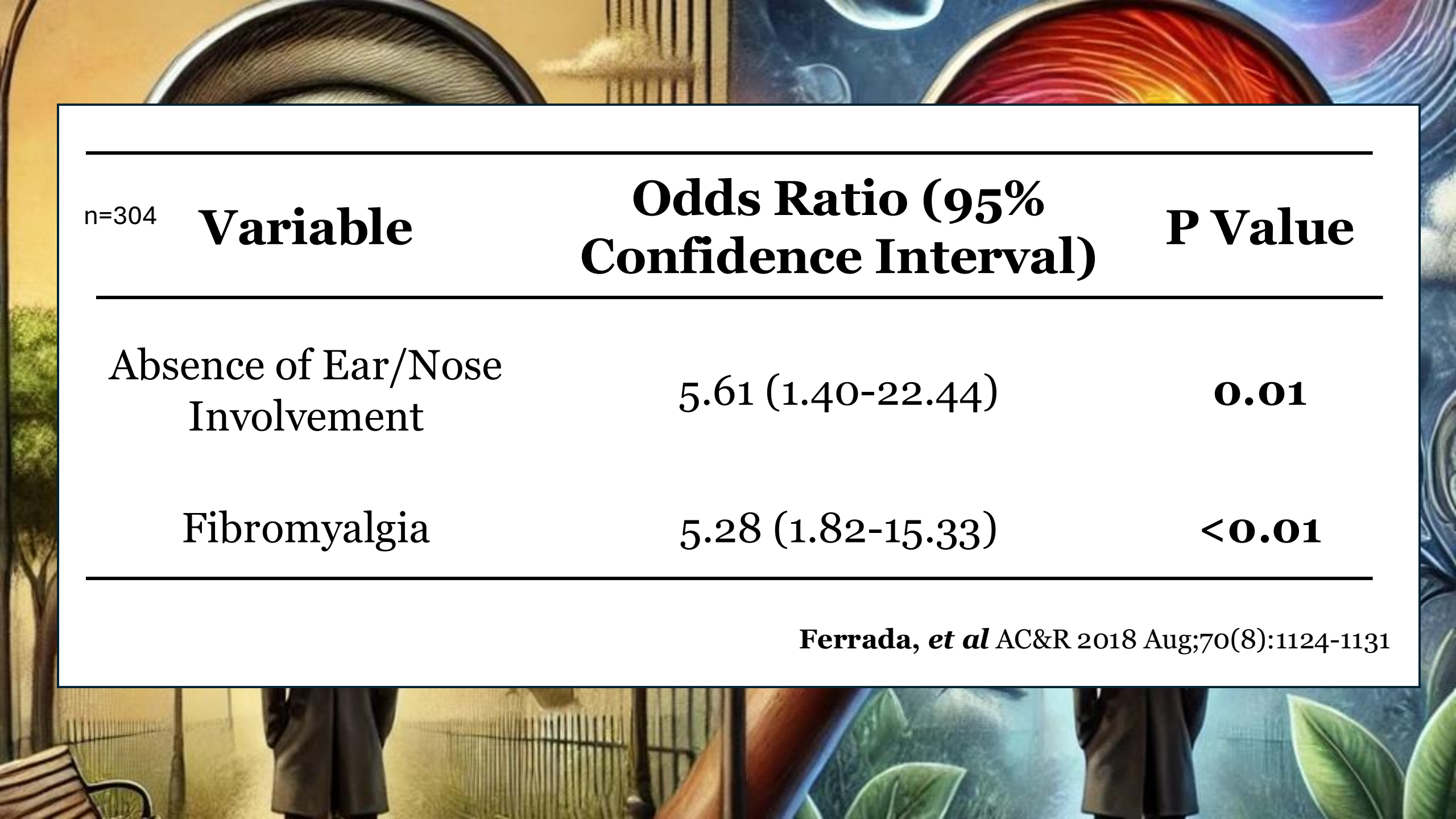
- **Lack clear definitions**
- **Have not been validated**
- **Small cohorts**



- **54% saw more than 3 physicians prior to establishing a diagnosis**
- **55% of patients went to the emergency room prior to diagnosis because of RP symptoms**

n=304

Ferrada, *et al* AC&R 2018 Aug;70(8):1124-1131



n=304 Variable	Odds Ratio (95% Confidence Interval)	P Value
Absence of Ear/Nose Involvement	5.61 (1.40-22.44)	**0.01**
Fibromyalgia	5.28 (1.82-15.33)	**<0.01**

Ferrada, *et al* AC&R 2018 Aug;70(8):1124-1131

Who wants to order Collagen II Antibodies?

Who wants to do a Biopsy?

Is it necessary for patients with RP to have elevated inflammatory markers to make a diagnosis?

Relapsing polychondritis

VERY

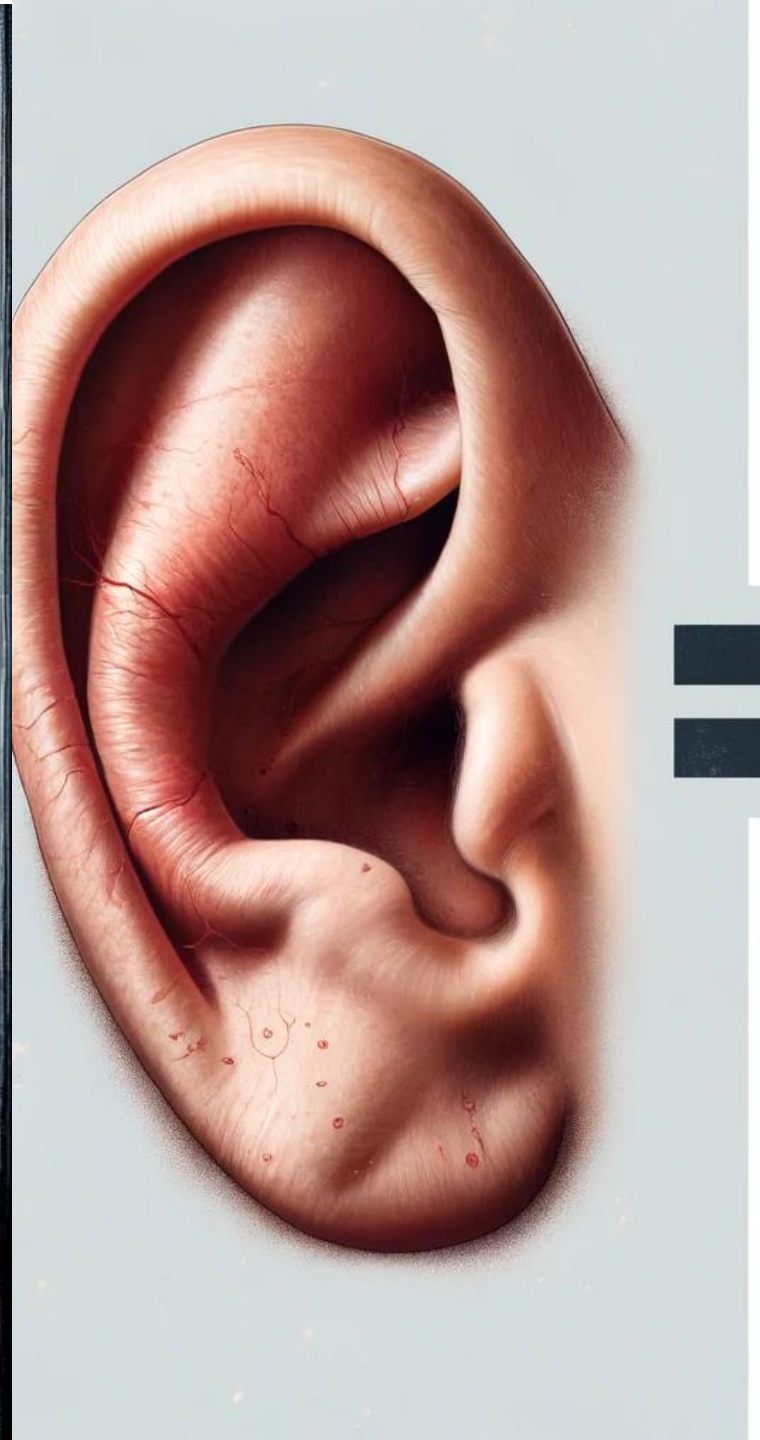
Hetereogenous disease

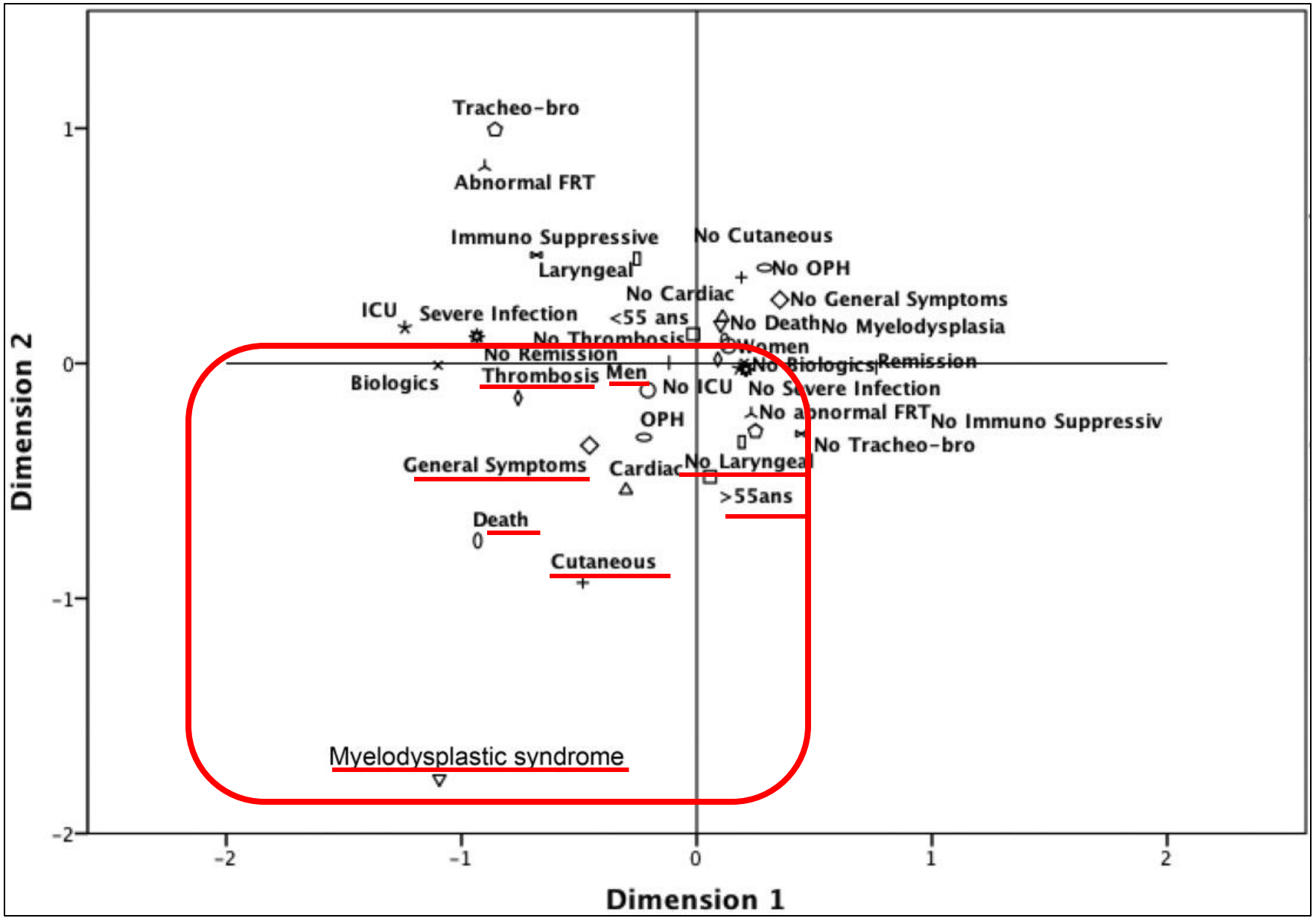
Multiple clinical manifestations

More than one disease

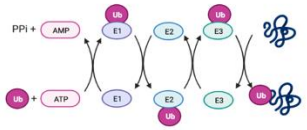
Diagnosis Pearls

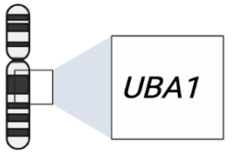
- Are all your sure all patients with fibromylagia dont have RP?
- No biomarker
- Inflammatory markers can be normal
- Do not forget about VEXAS!!

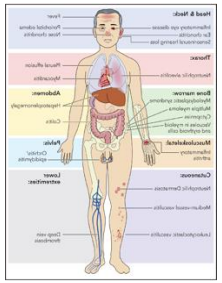


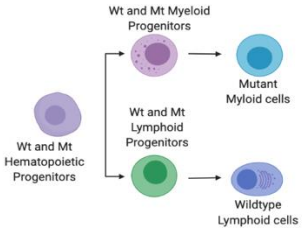


VEXAS





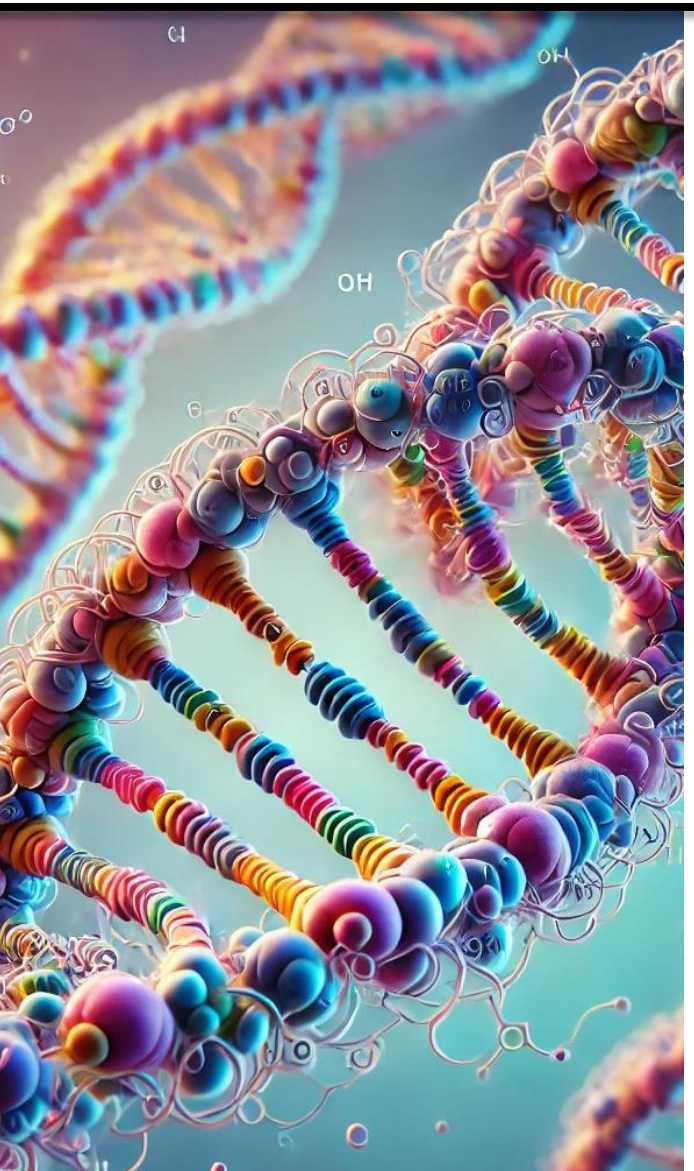




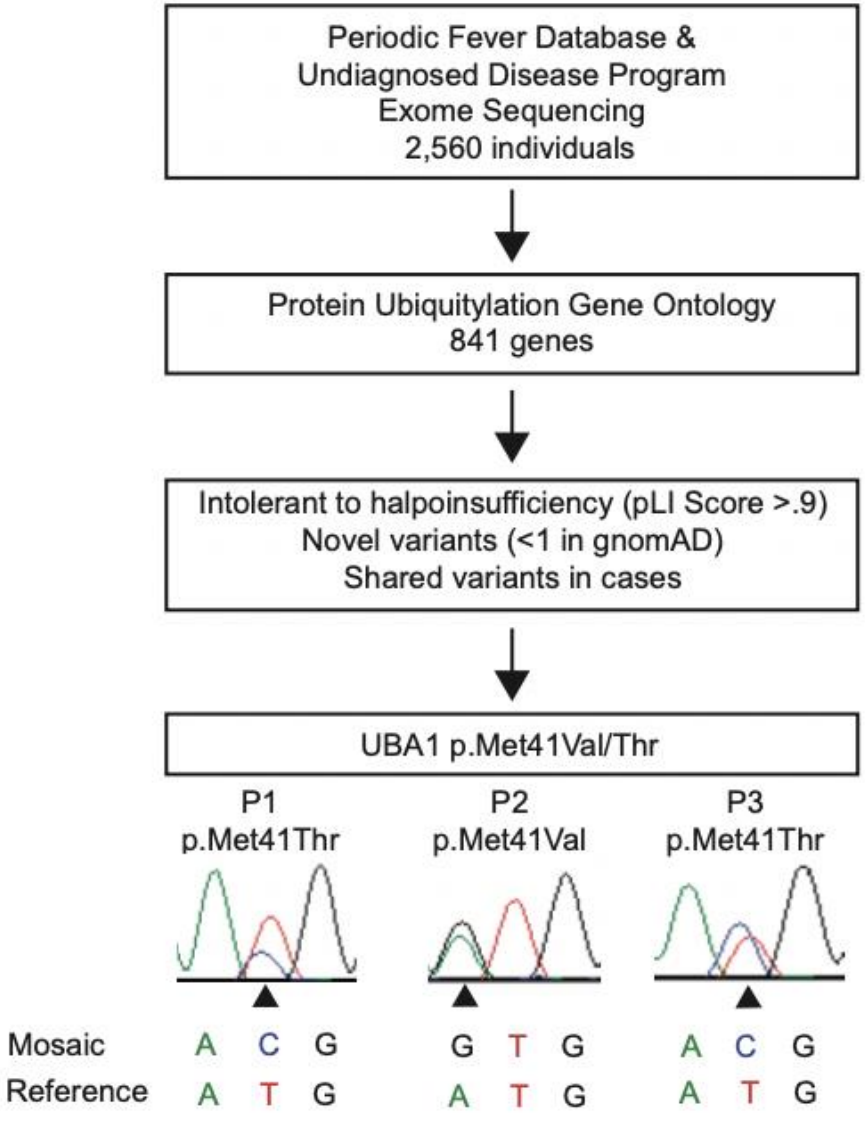
Dion et al, Arthritis Rheumatol 2016 Dec;68(12):2992-3001

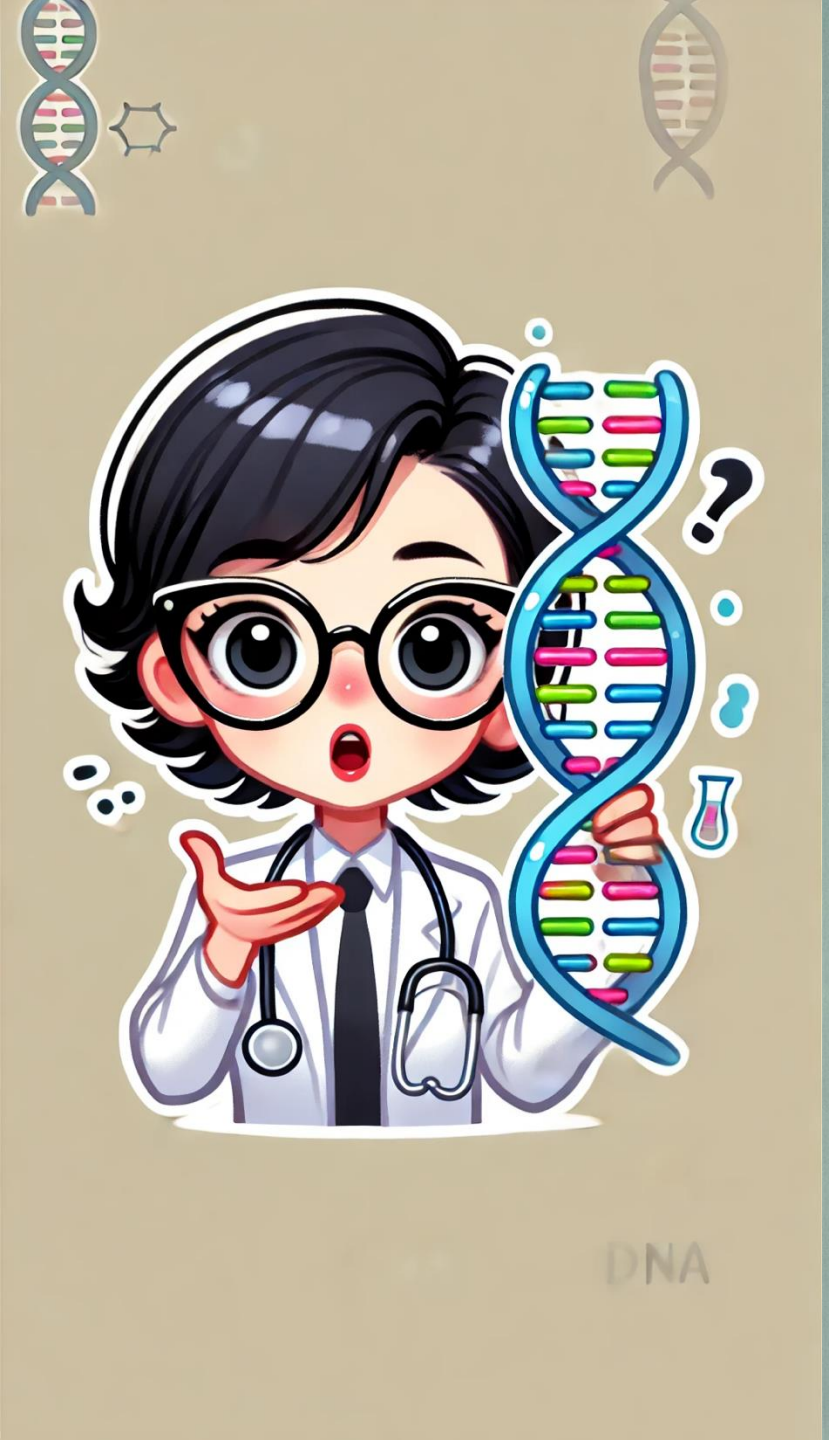
Beck et al, N Engl J Med. 2020 Dec 31;383(27):2628-2638

Genotype First Approach



David Beck

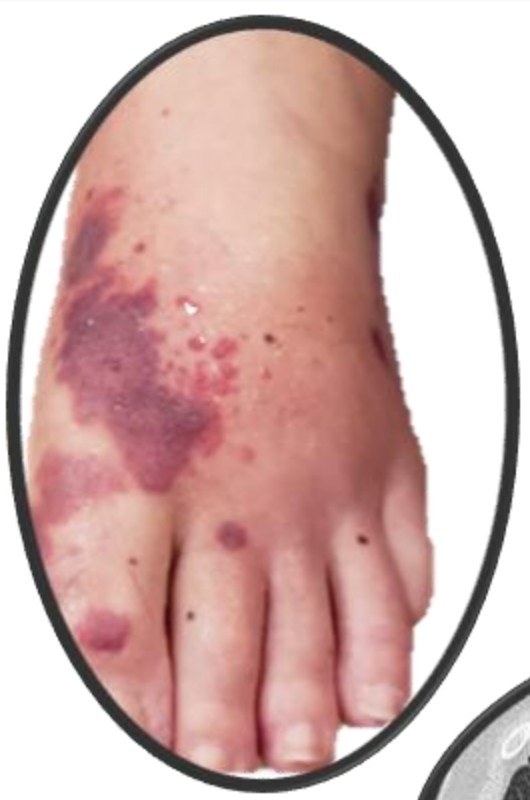


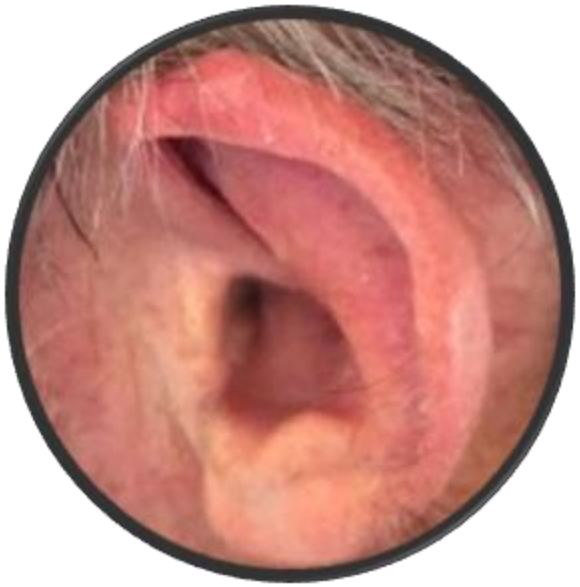

DNA

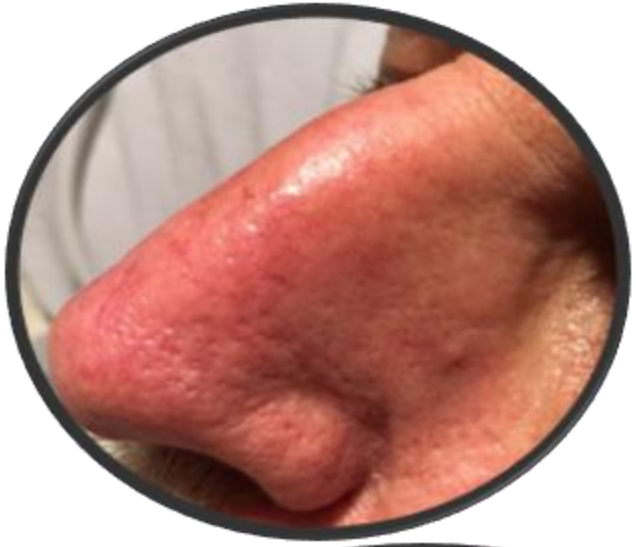

E1
ENZYME
TICKET
UBIQUTIN

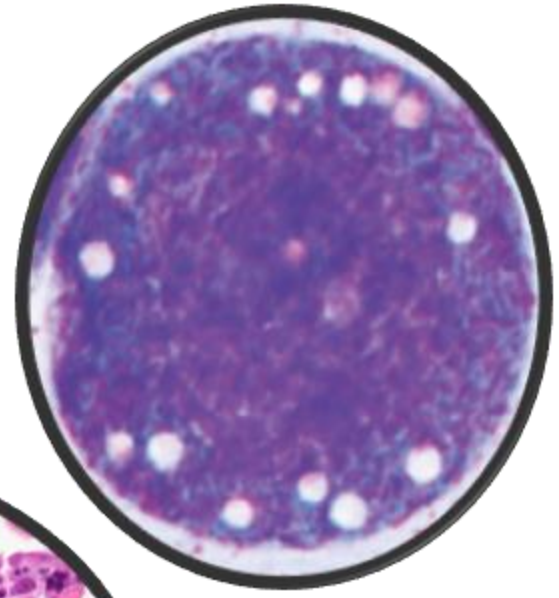

UBIQUTIN
UBIQUTIN

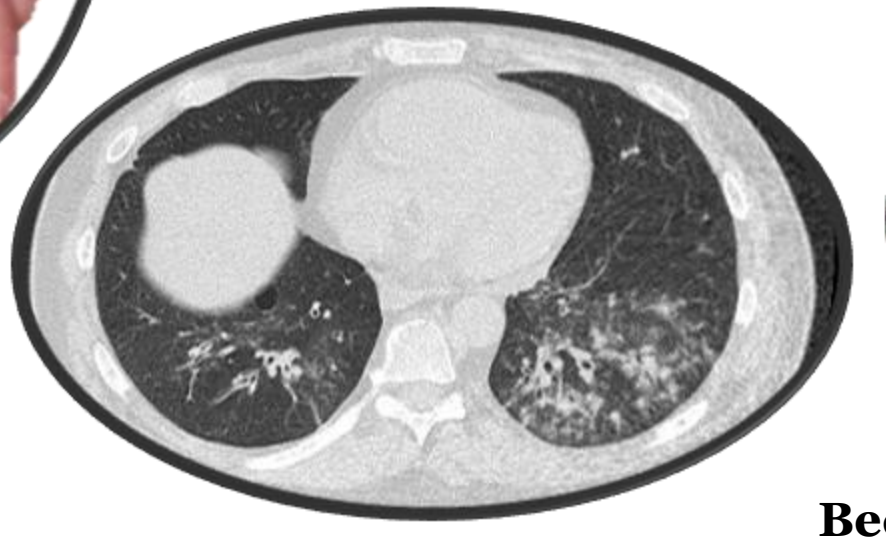
Clinical Manifestations in VEXAS

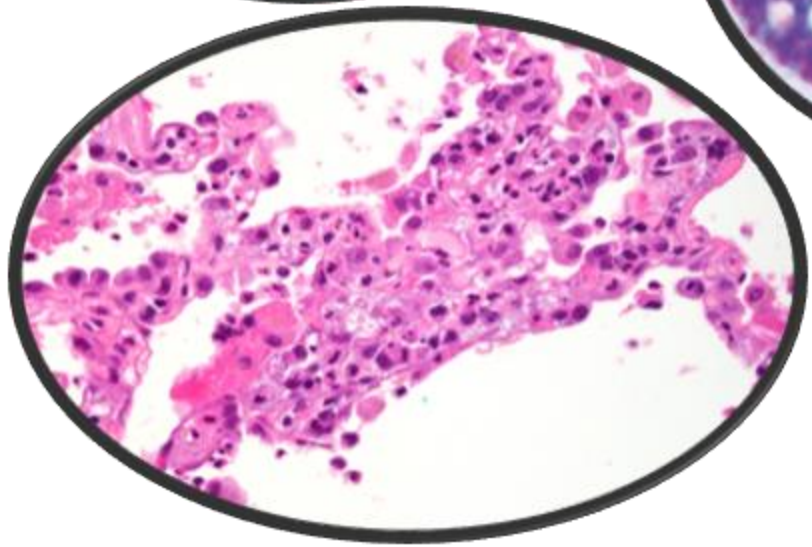






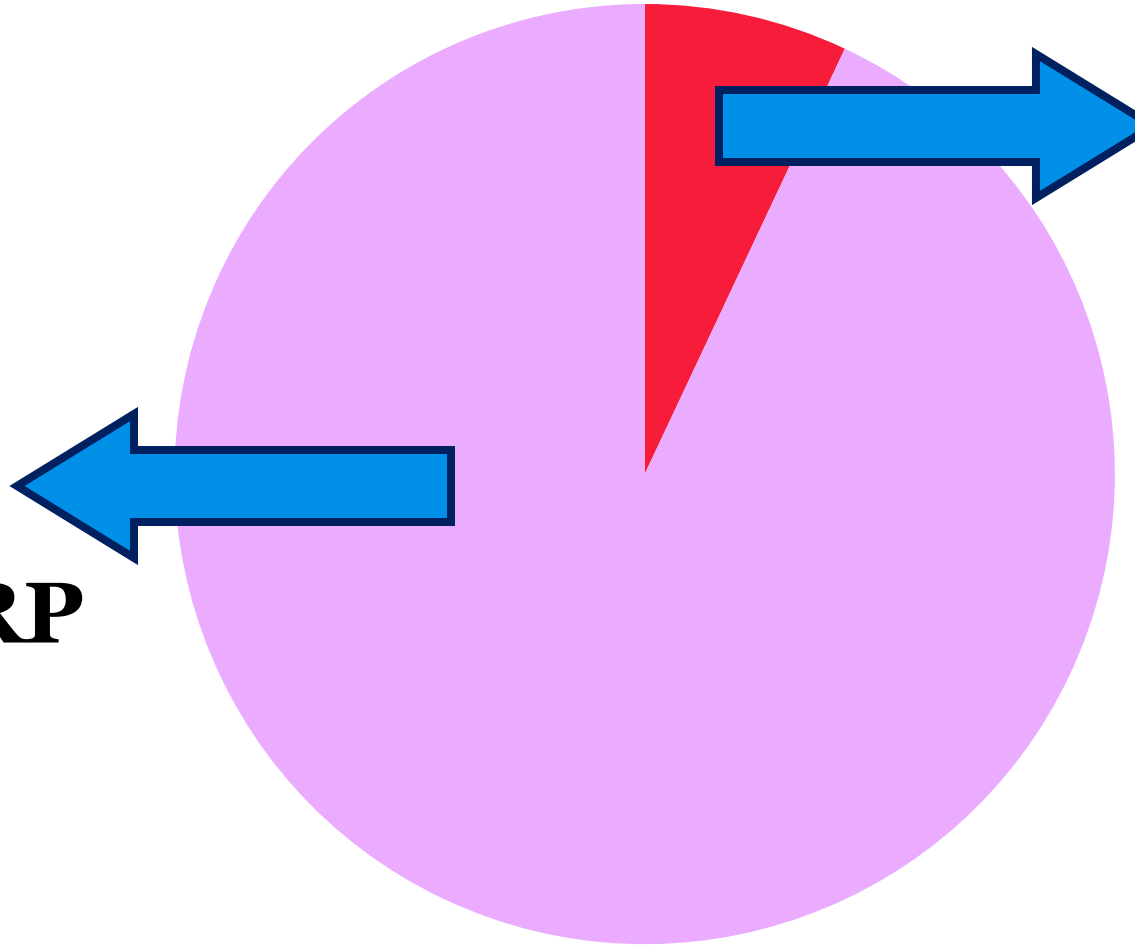






Beck et al, N Engl J Med.2020 Dec 31;383(27):2628-2638

8% of Patients with RP have VEXAS



n= 92
Patients with RP

n=7
8% VEXAS-RP

Ferrada, et al Arthritis Rheumatol. 2021 Mar 28.

Patients with VEXAS-RP vs RP

- Have distinct characteristics
  - Older
  - Skin lesions
  - Pulmonary infiltrates
  - Heart disease
  - Eye inflammation
  - Pulmonary infiltrates
  - **Airway disease???**
- Have more complications
  - Increased on mortality
  - ICU admissions
  - Diagnosis of MDS
  - Refractory disease

Tschida N, et al. Ann Rheum Dis. 2021 Aug:80(8):1057-1061
Khitri et al RMD Open. 2022 Jul;8(2):e002255
Ferrada et al Arthritis Rheumatol. 2021 Oct;73(10

Patients with VEXAS-RP Can be Identified Using Clinical Variables

A male patient with ear and nose chondritis…

- MCV >100 or
- Platelets <200

100 % Sensitivity
96 % Specificity

Ferrada et al Arthritis Rheumatol. 2021 Oct;73(10):1886-1895

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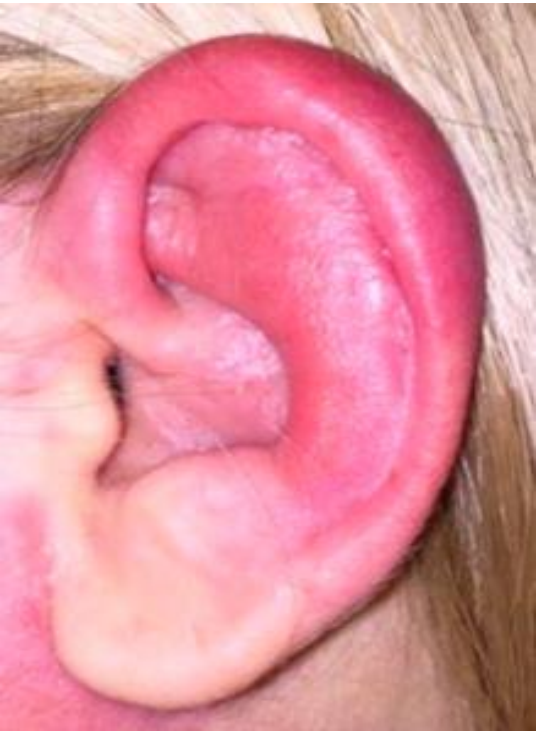
a S

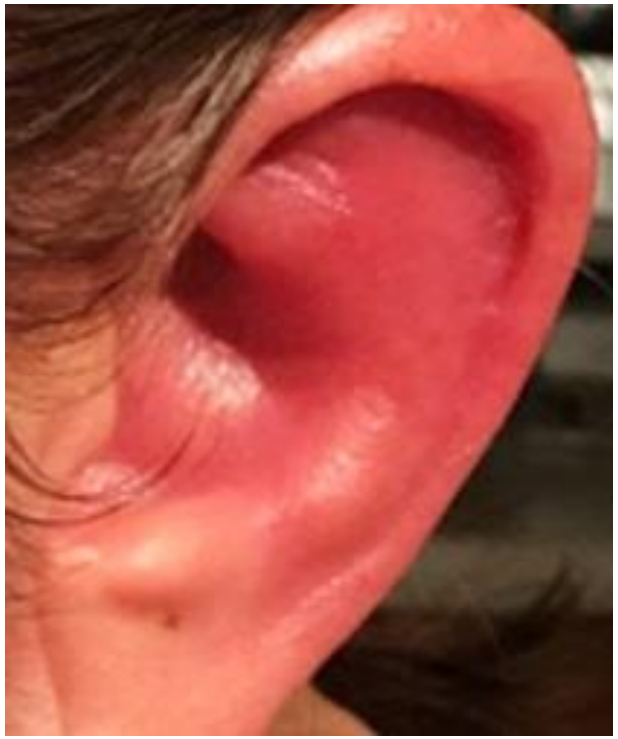

IMPORTANT
WE AGREE!
WE AGREE!
RESPECT THE EAR!
LEADER OF LISTENING!
WE AGREE!

Which one is consistent with RP?

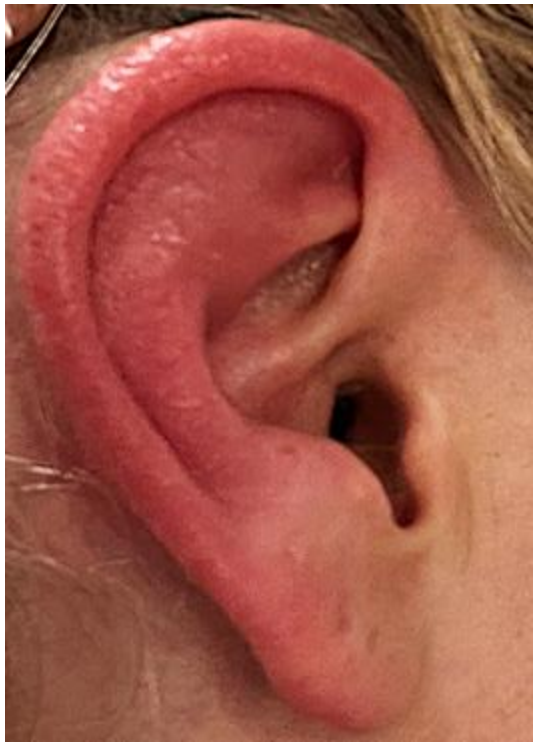
A.



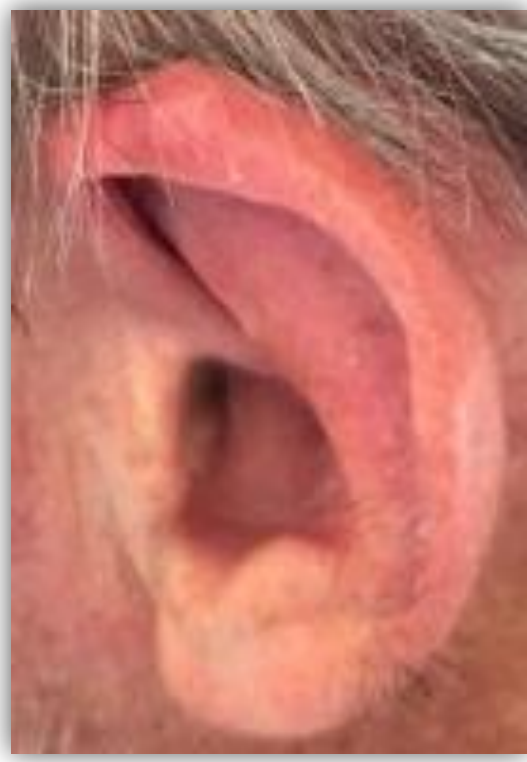
B.



C.



D.



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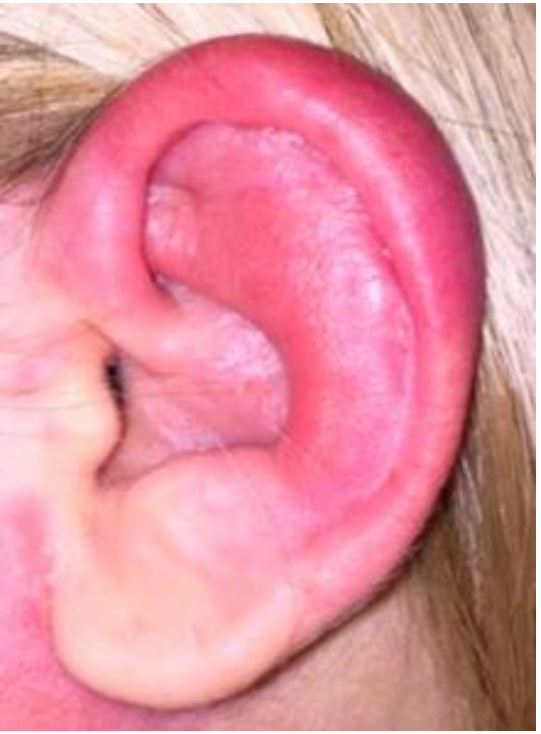
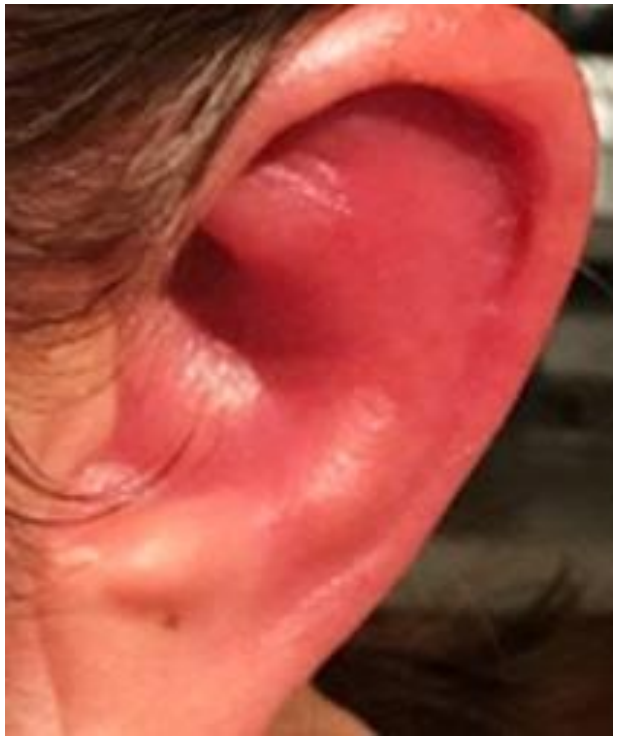


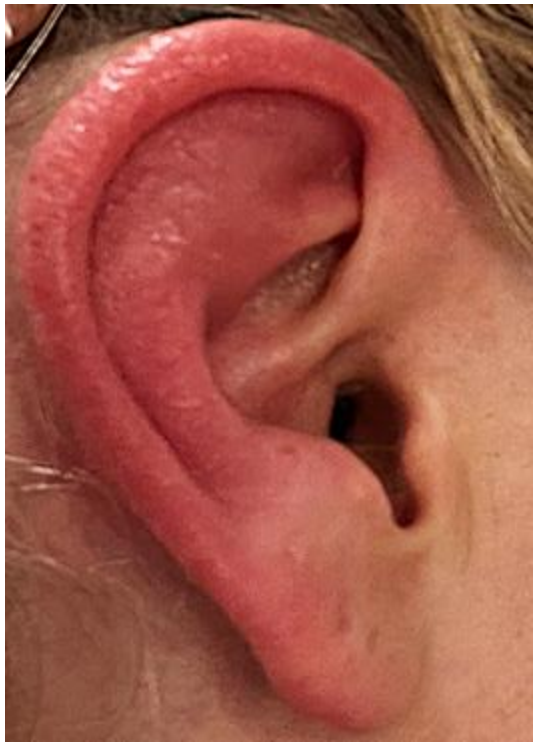
RP

B.



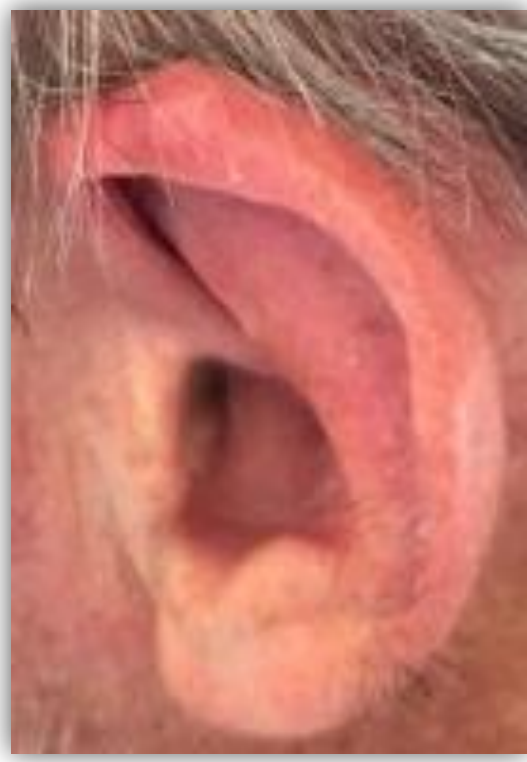
GPA

C.

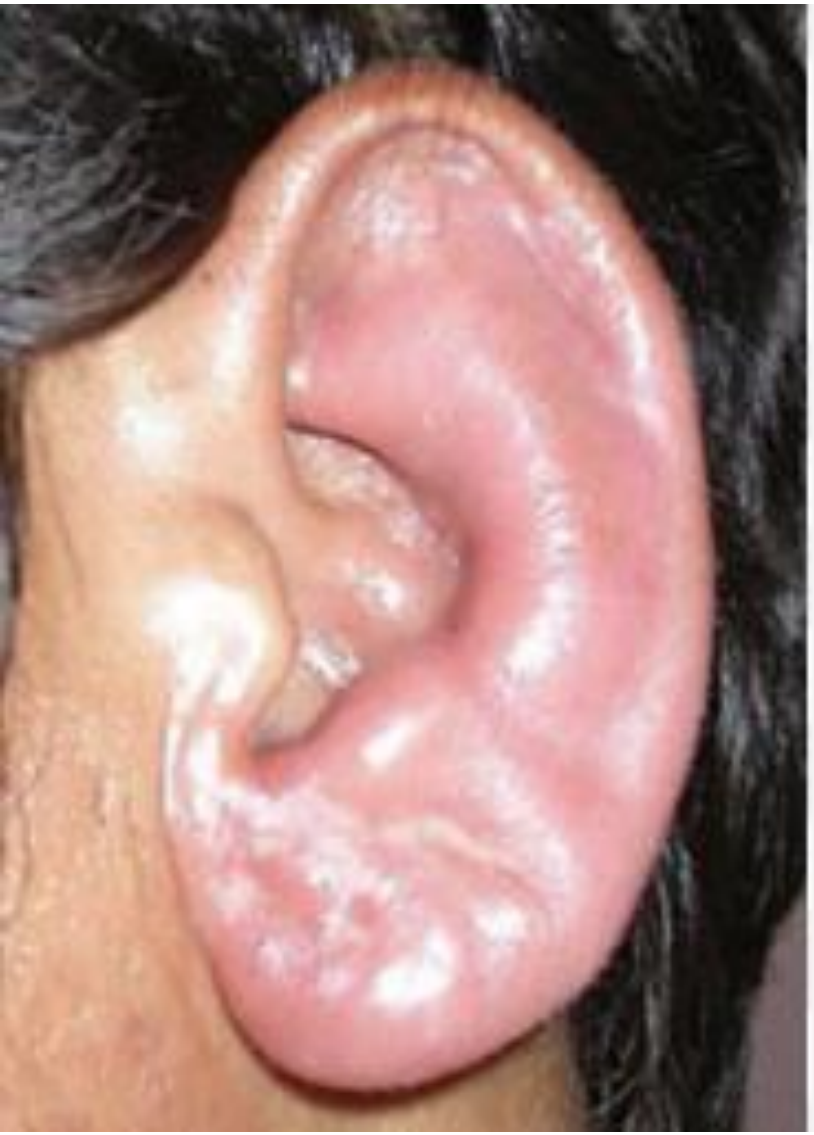


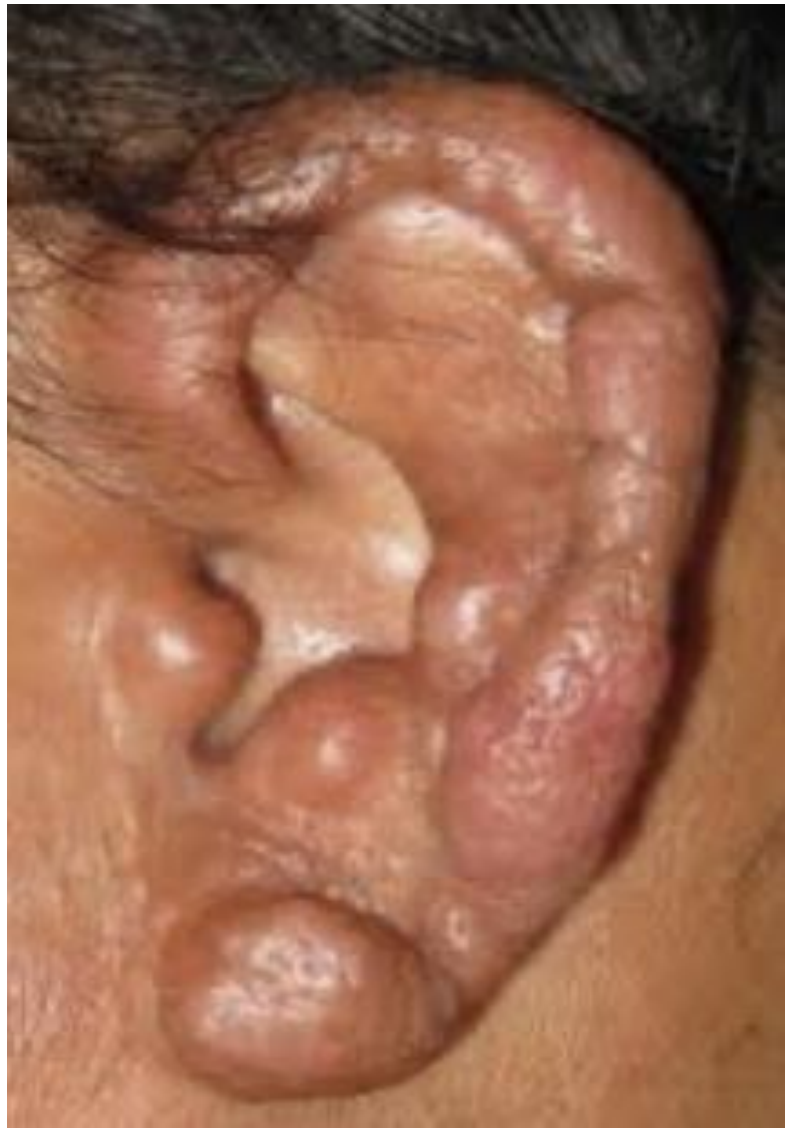
Psoriasis

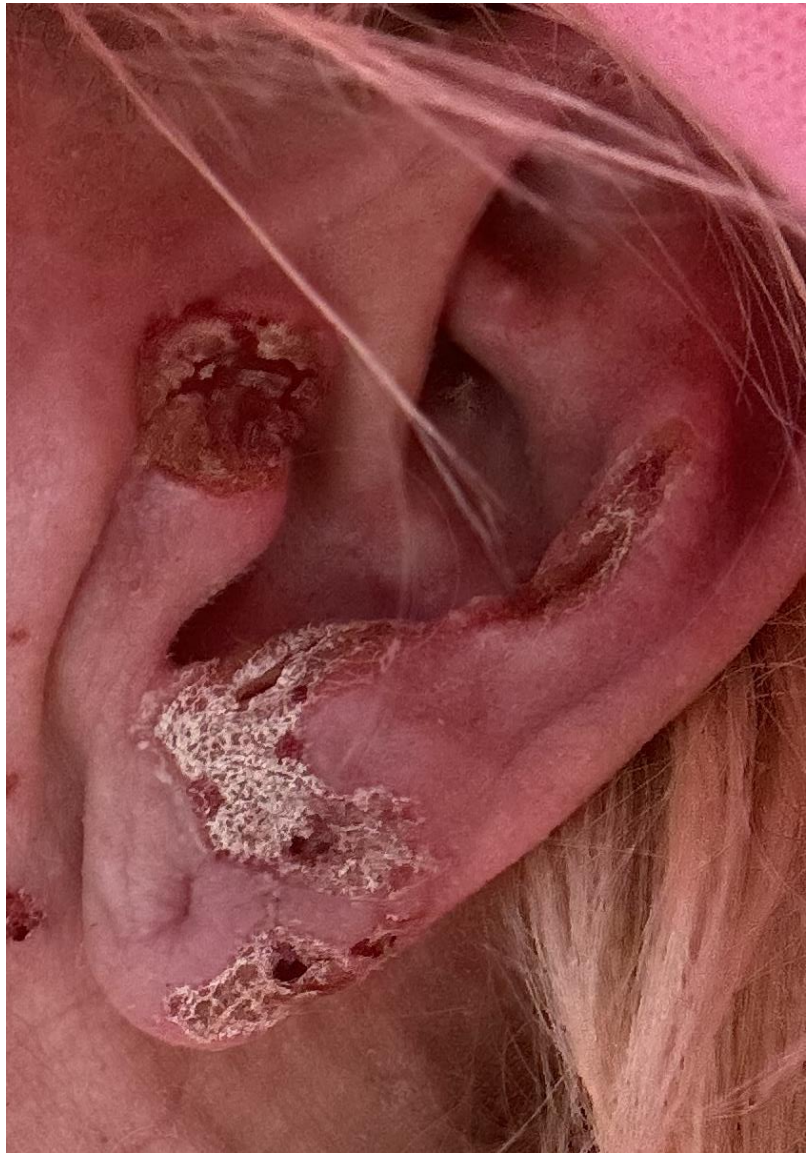
D.



VEXAS

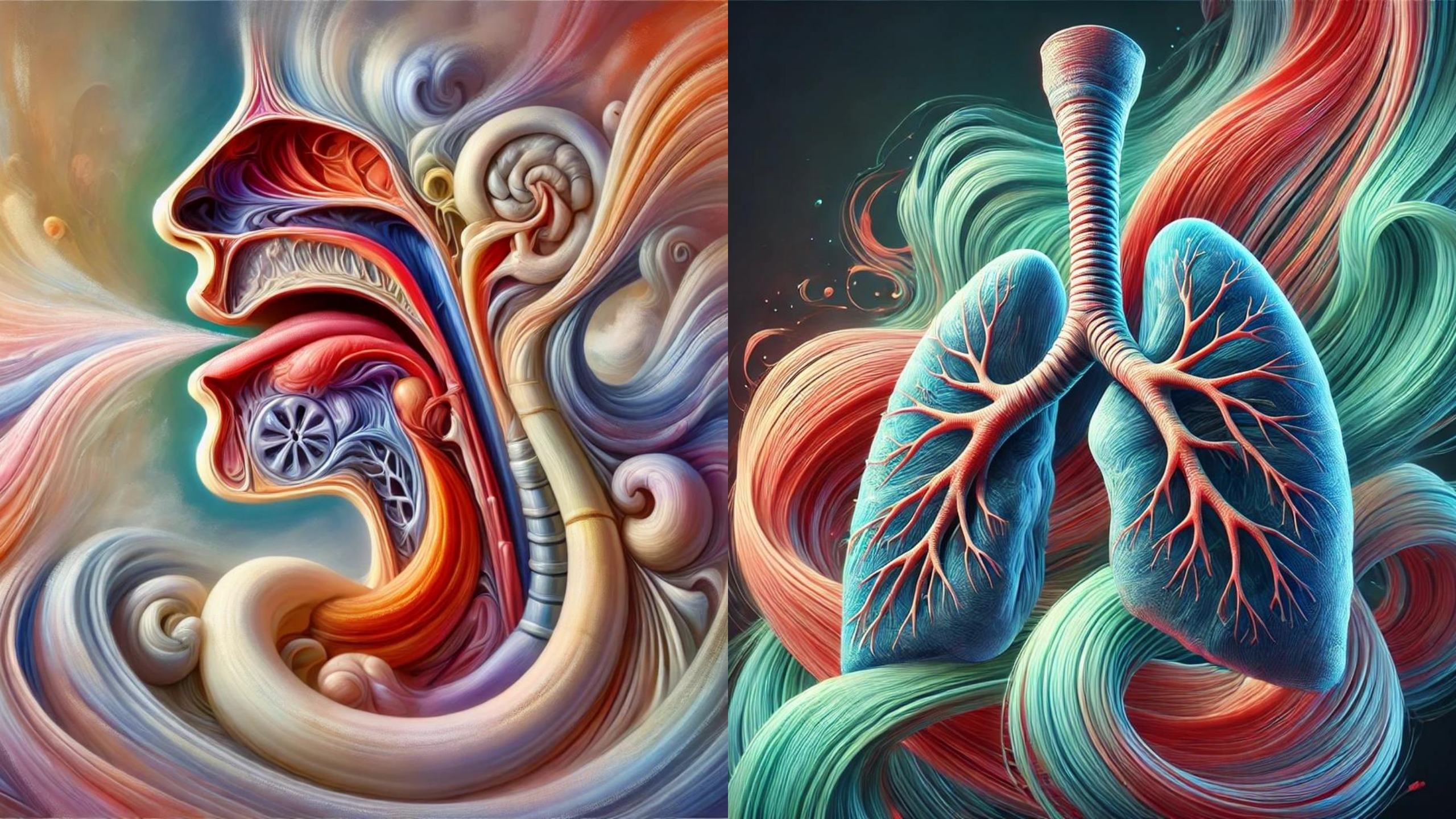




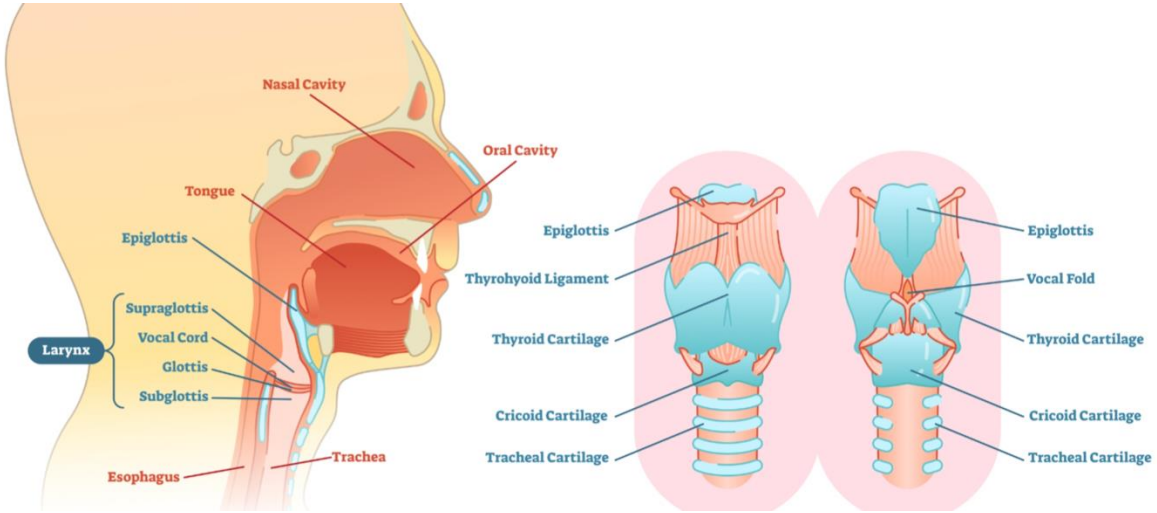


Ear Pearls

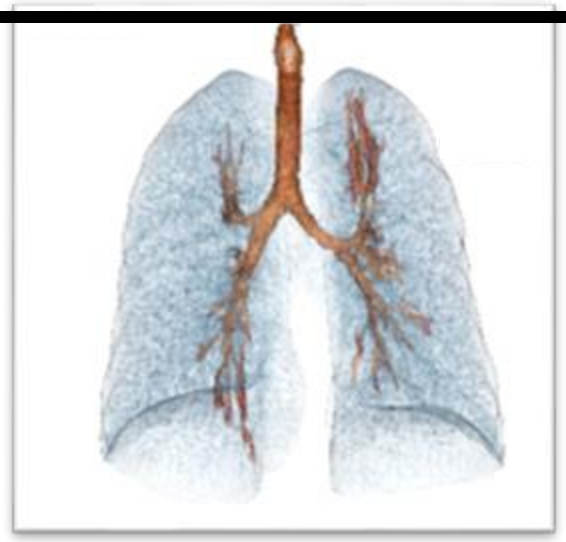
- Most physicians only diagnosed RP when there is ear chonritis
- Ear involvement may arise later in the disease course
- Clinical presentation of ear symptoms is heterogeneous and may progress over time.
- Cauliflower ear is not necessary for diagnosis
- **Not all red ears are due to RP**
- **Associated Clinical Features and history is the key!!**

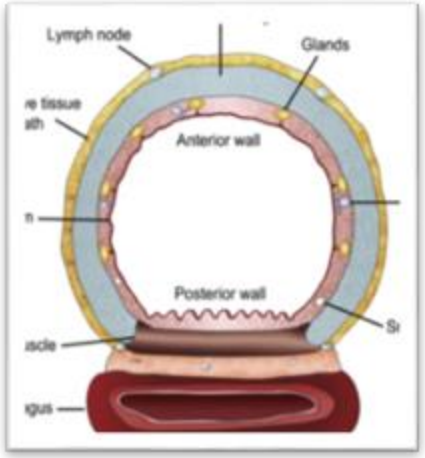


Upper Airway

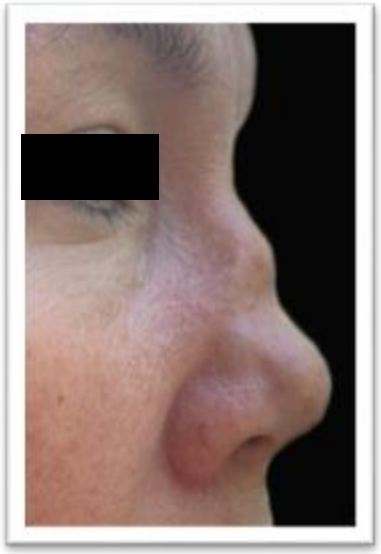


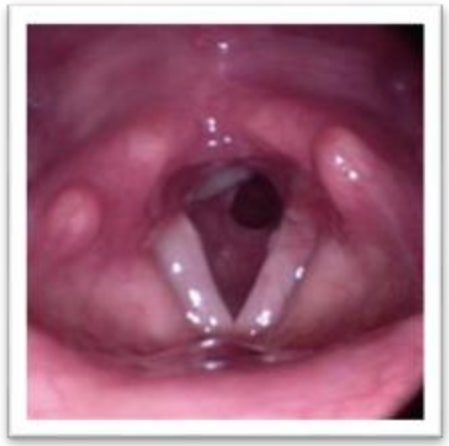
Lower Airway

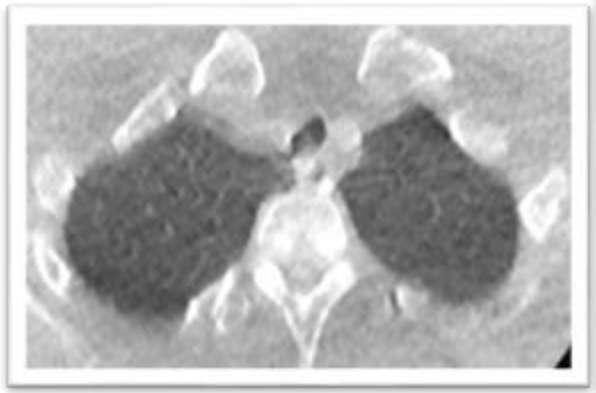


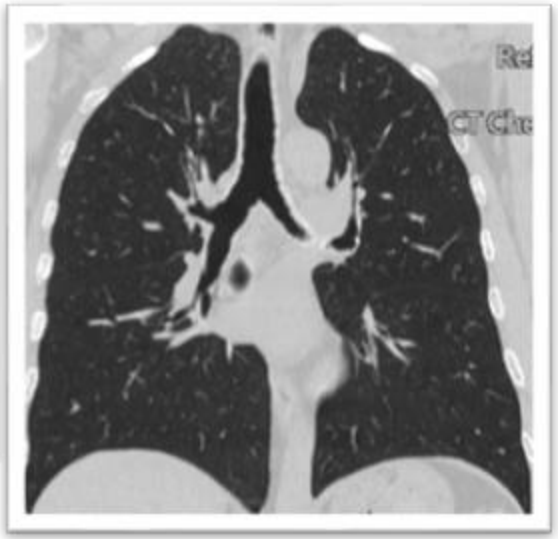


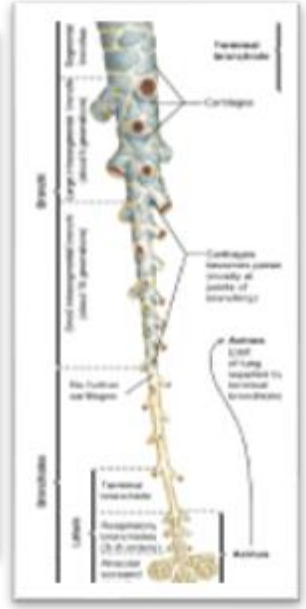
https://medictests.com/units/basic-airway-anatomy#images-3

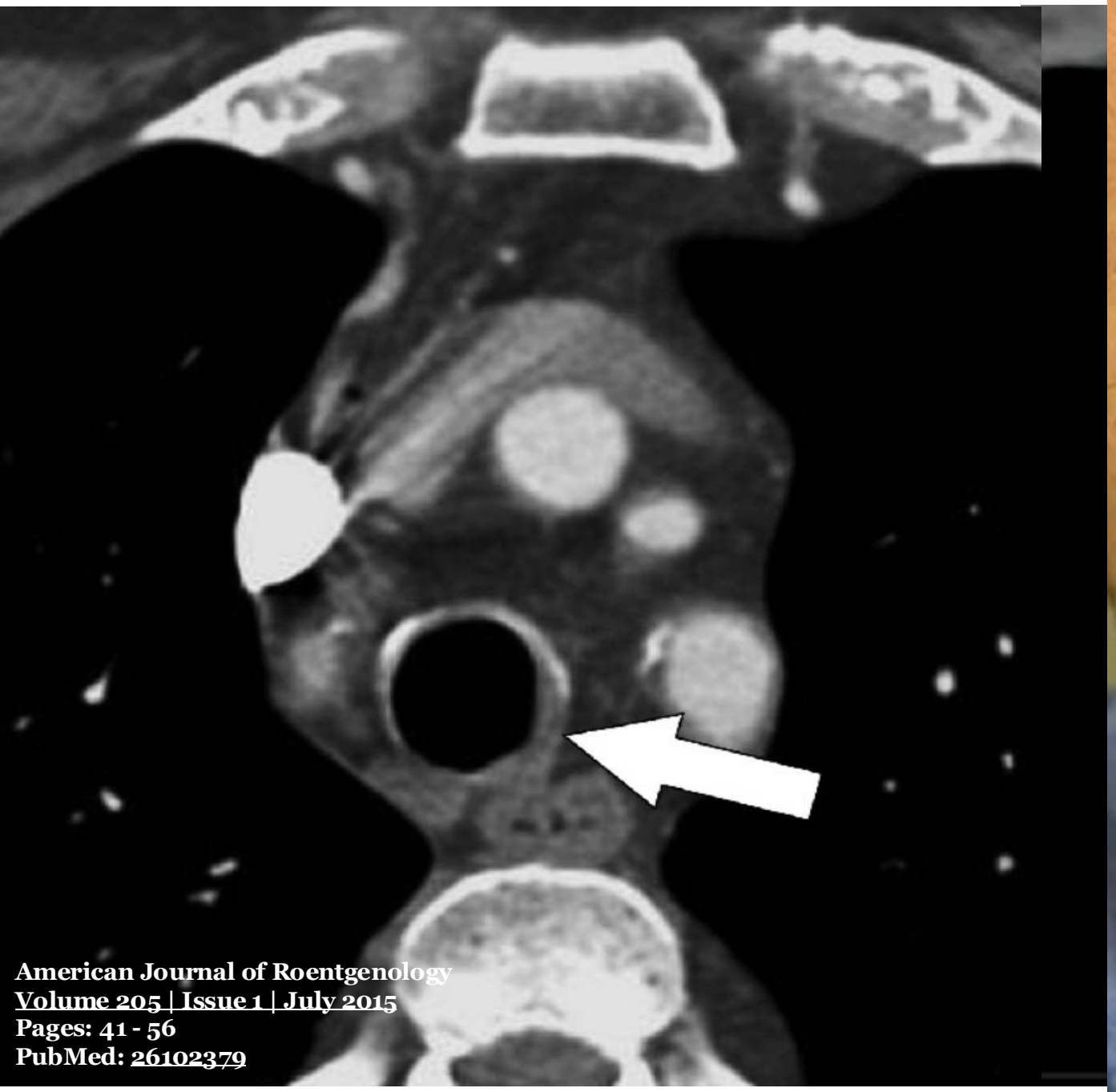




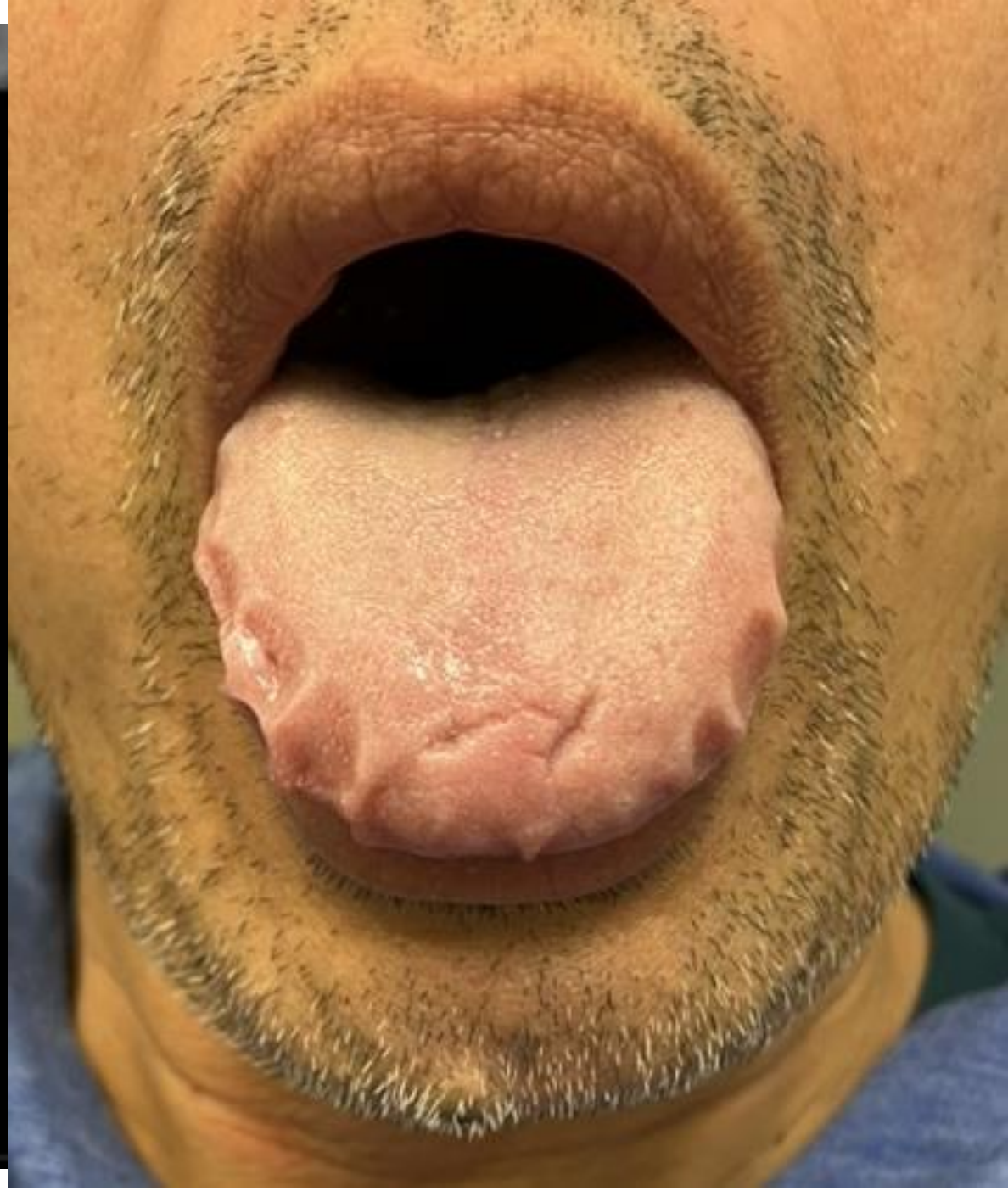


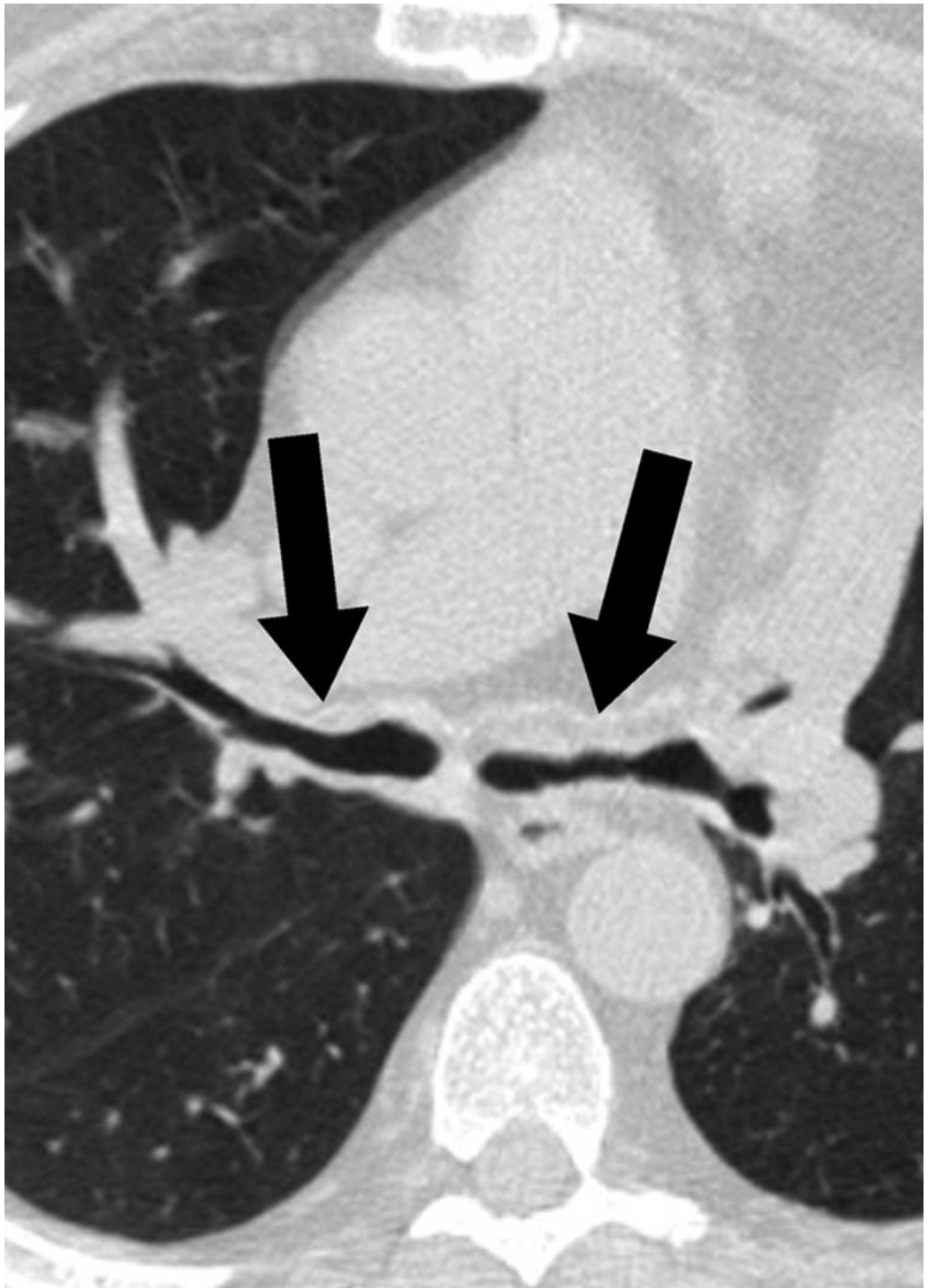


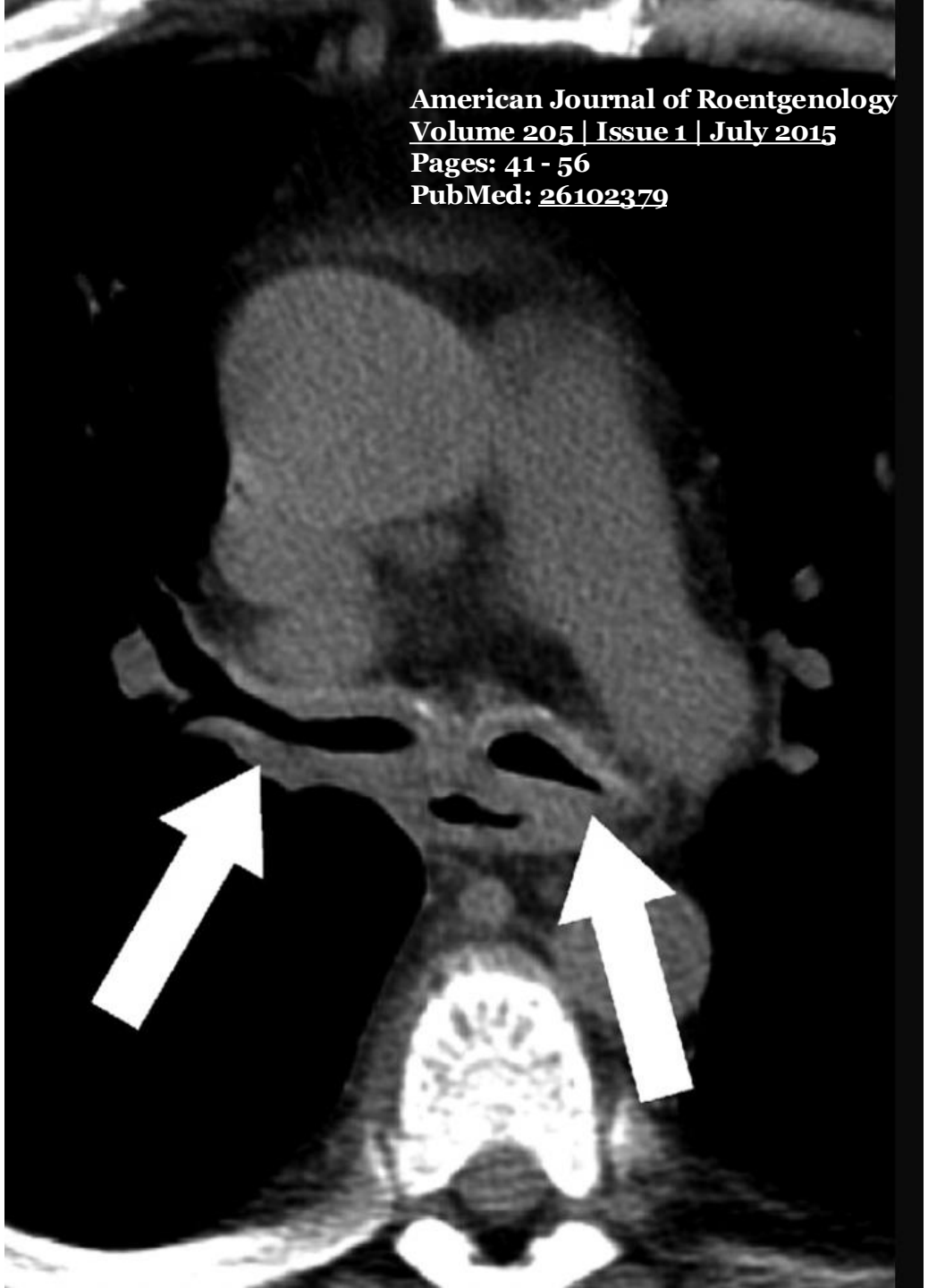




American Journal of Roentgenology
Volume 205 | Issue 1 | July 2015
Pages: 41 - 56
PubMed: 26102379







American Journal of Roentgenology
Volume 205 | Issue 1 | July 2015
Pages: 41 - 56
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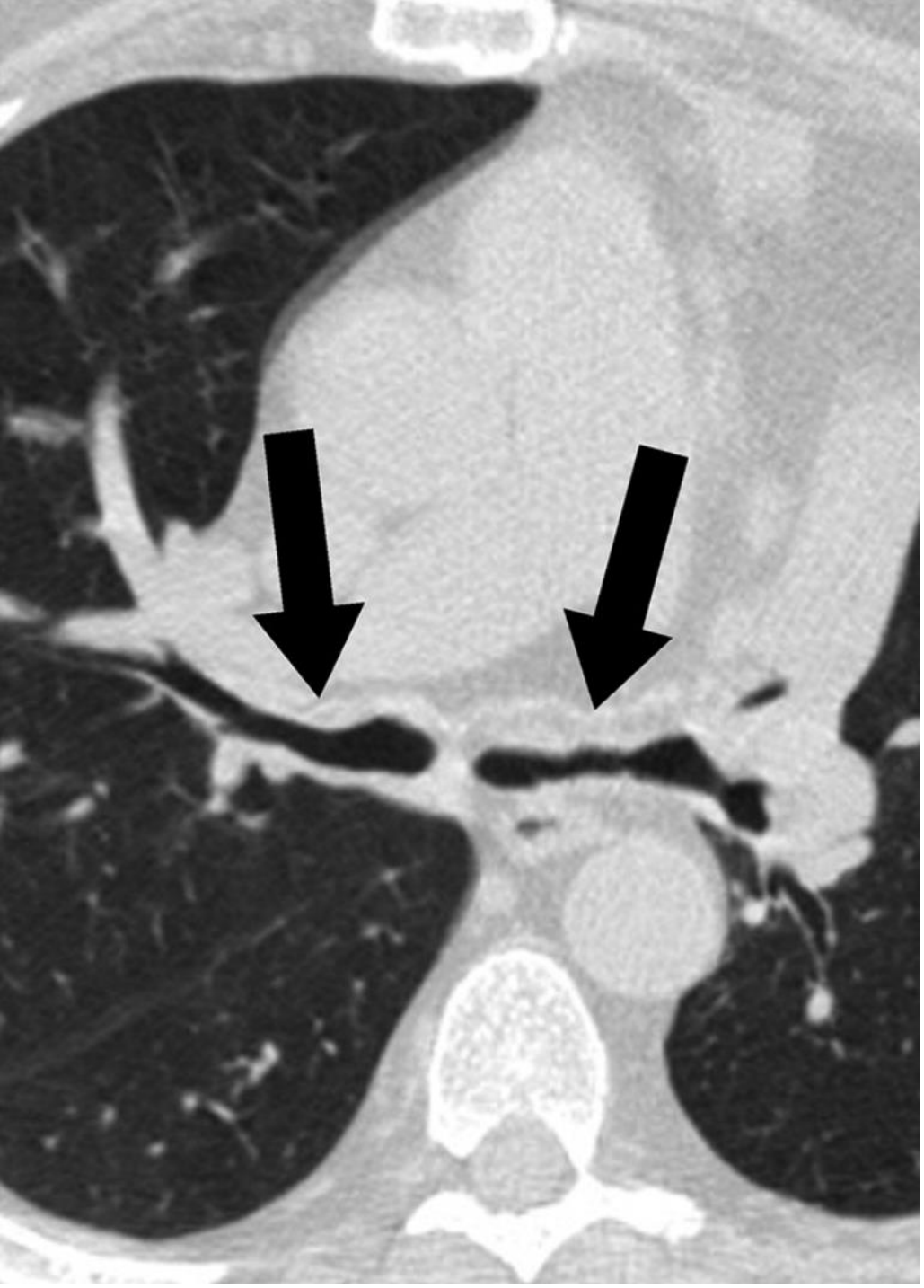
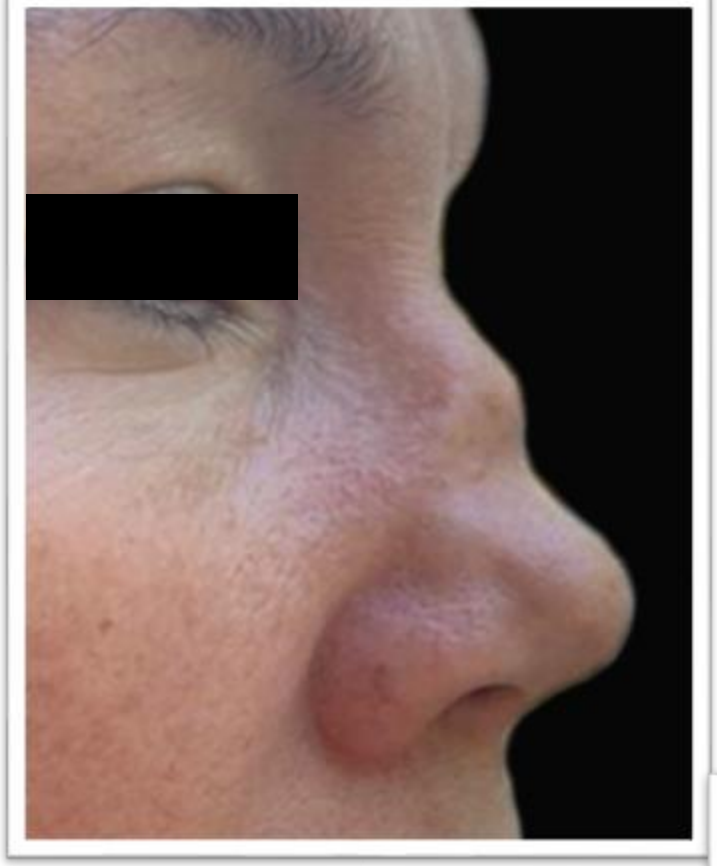
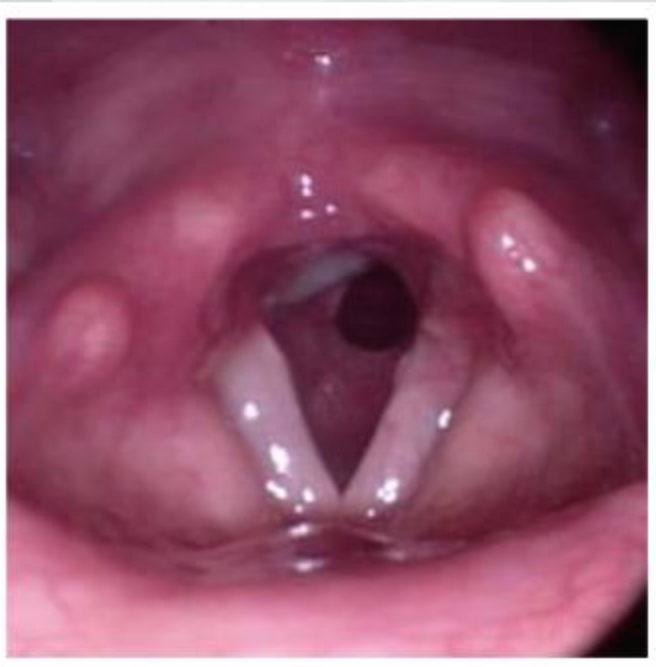
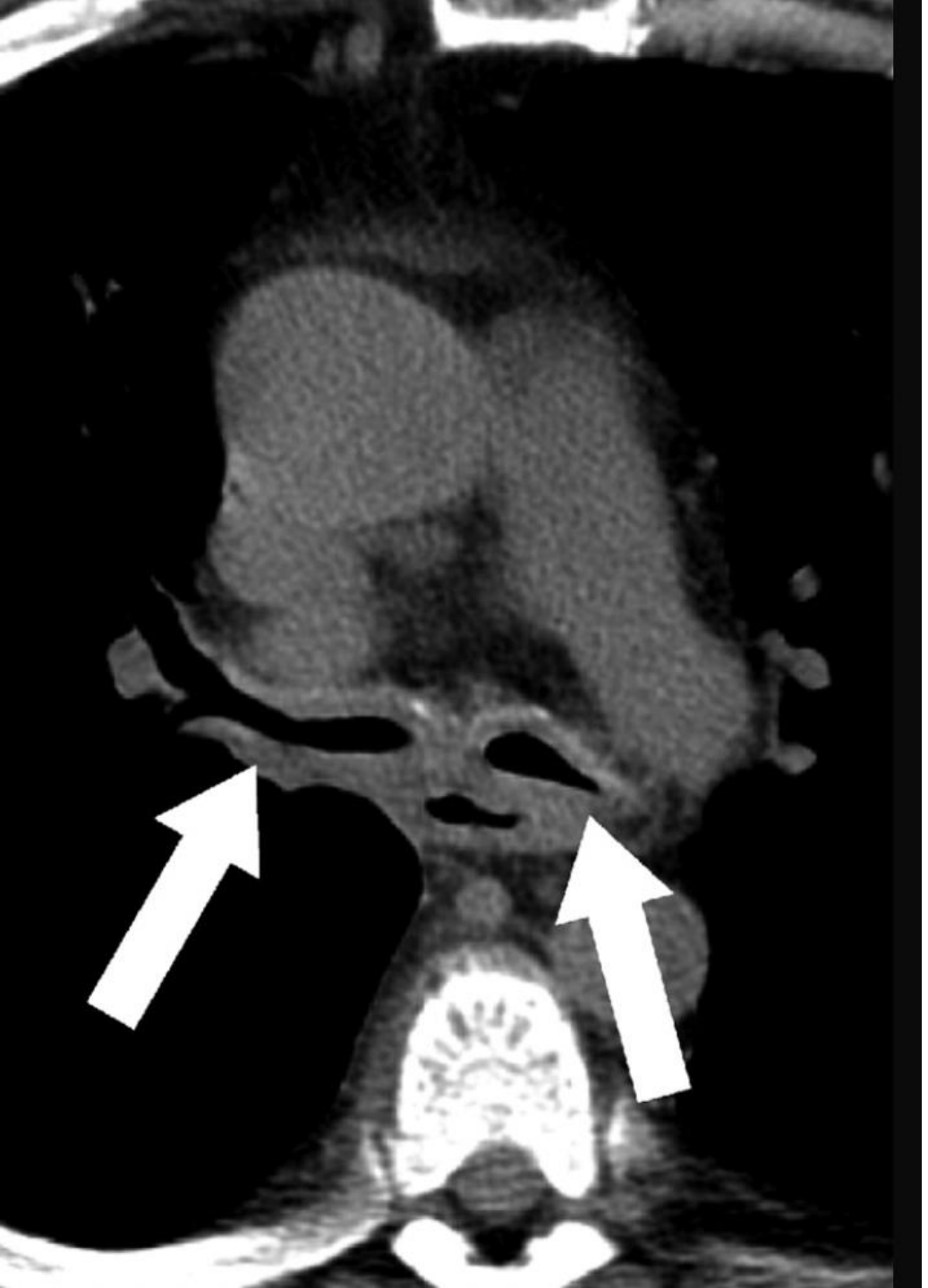
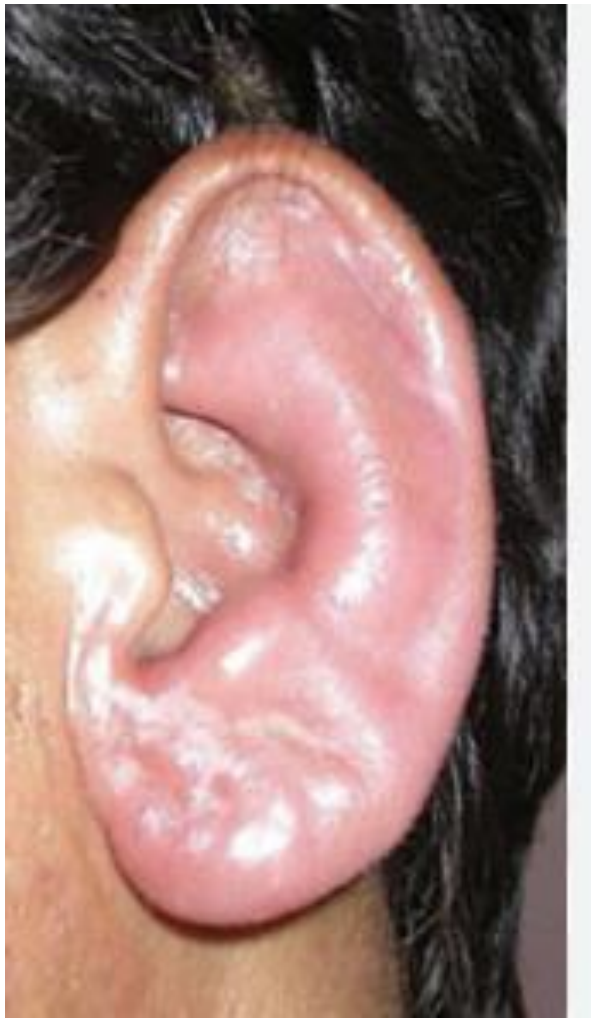
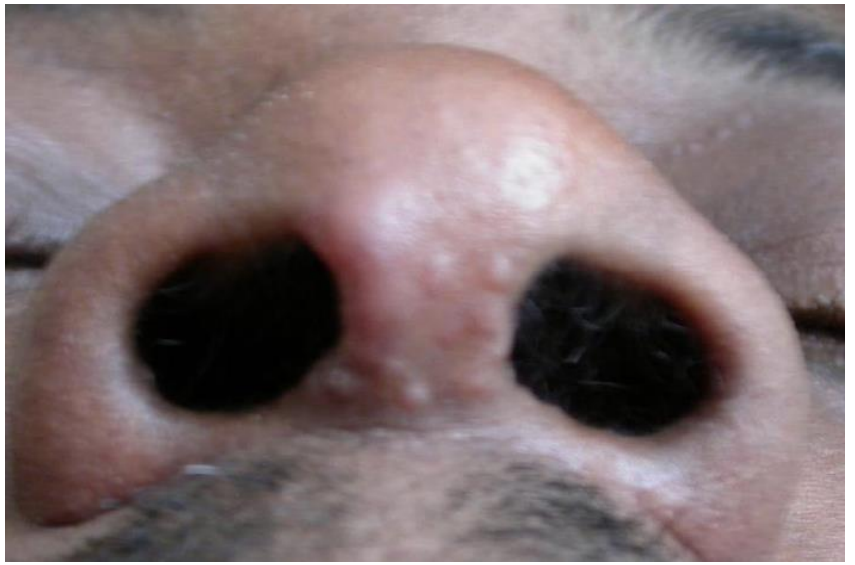


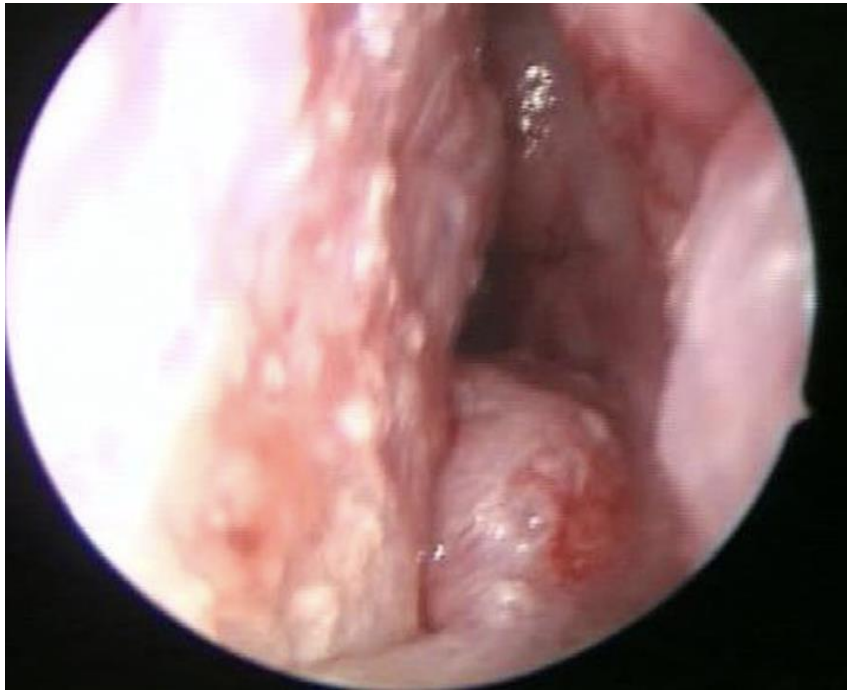




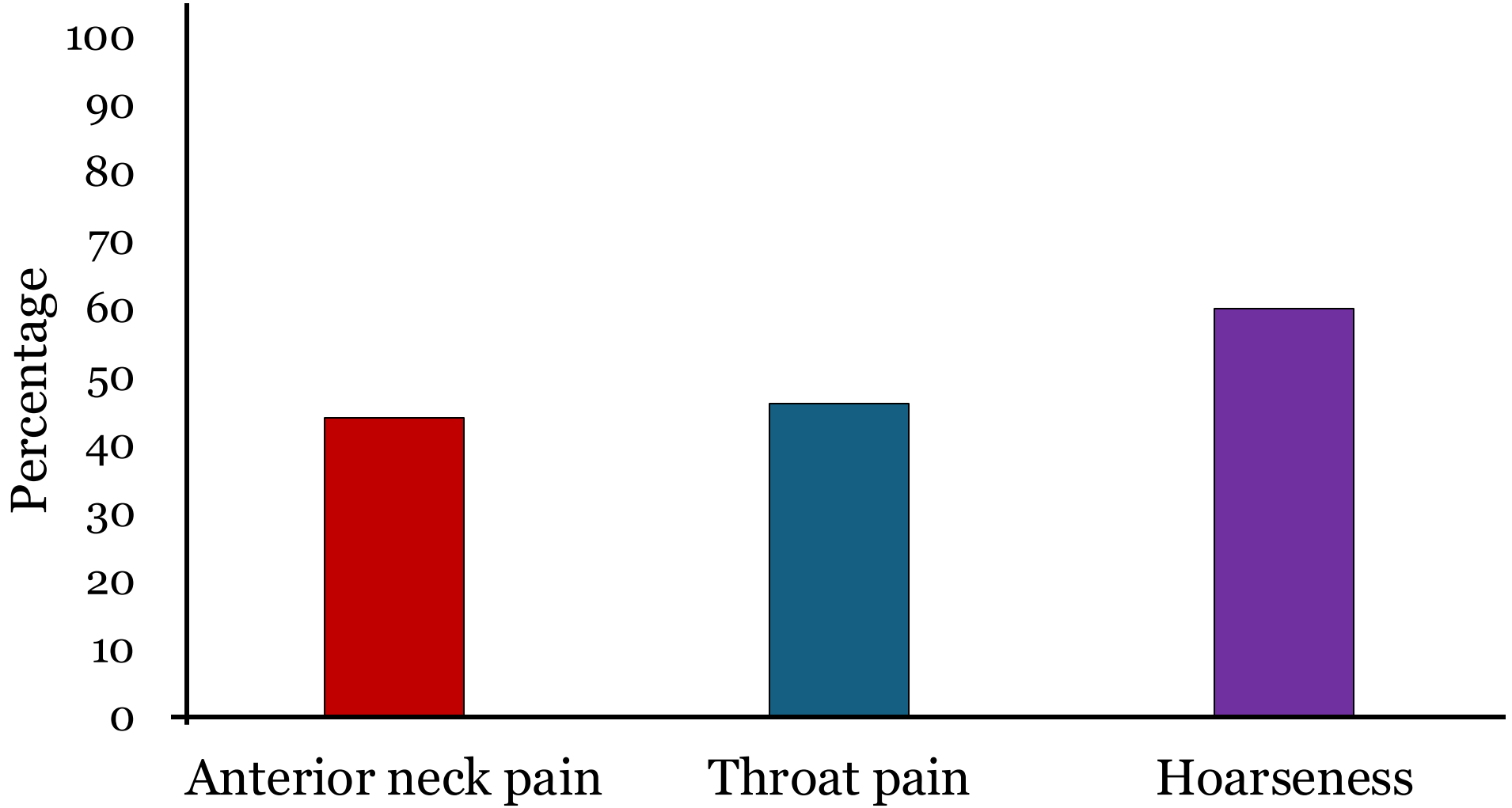








Common Neck Symptoms in PATIENTS WITH Relapsing Polychondritis

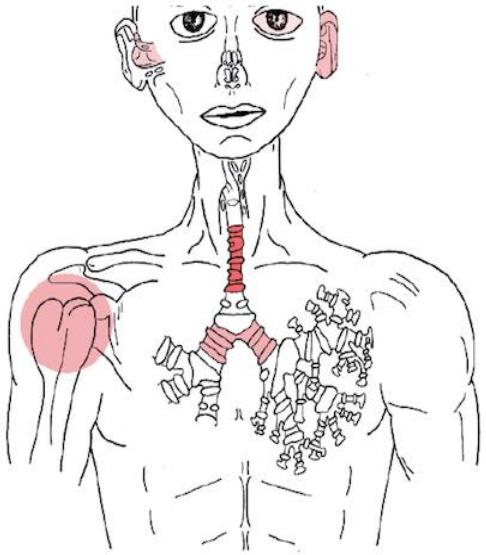


n=668

Ferrada Unpublished data

Airway Involvement in RP

- Prevalence 20 - 50 %
- Airway symptoms can be the first manifestation of the disease
  - Cough, stridor, hoarseness
- Delay in diagnosis
- Younger- more ICU admissions
- Have a previous diagnosis of asthma
- Higher incidence of costochondritis
- Less incidence of eye disease and auricular chondritis
- Complications
  - Subglottic stenosis
  - Focal and diffuse malacia



Type II

Ferrada, *et al* AC&R 2018 Aug;70(8):1124-1131
Dion *et al* Arthritis Rheumatol 2016 Dec;68(12):2992-3001
Wang, et al BMC Puml Med 2022 Jun 8;22(1):222

How do you recognize airway involvement?

Upper airway

- Cauliflower ear
- Saddle nose deformity
- Stridor

Lower airway

- Adult onset “asthma”
- Oral and genital ulcers (MAGIC)
- Wheezing

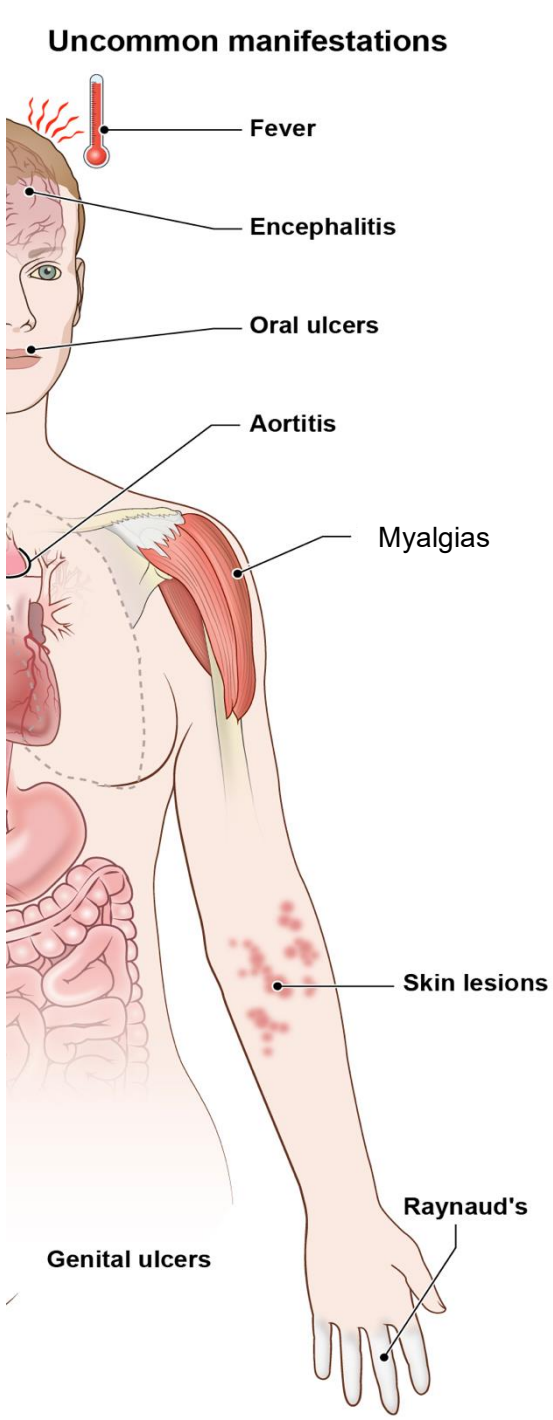
What do you do?

- E
- L

*These are the patients who will likely **NEED** acute treatment with corticosteroids*

- **Bronchoscopy**

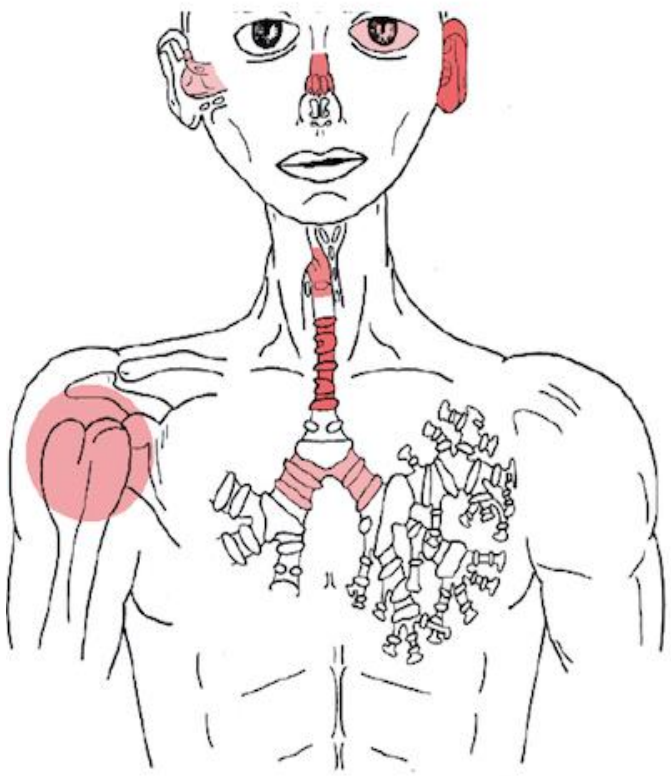
1. Think Airway Early
2. Know the Anatomy
3. Investigate Properly
4. Multidisciplinary Is Essential
5. Treat Aggressively – when?
6. Monitor Closely



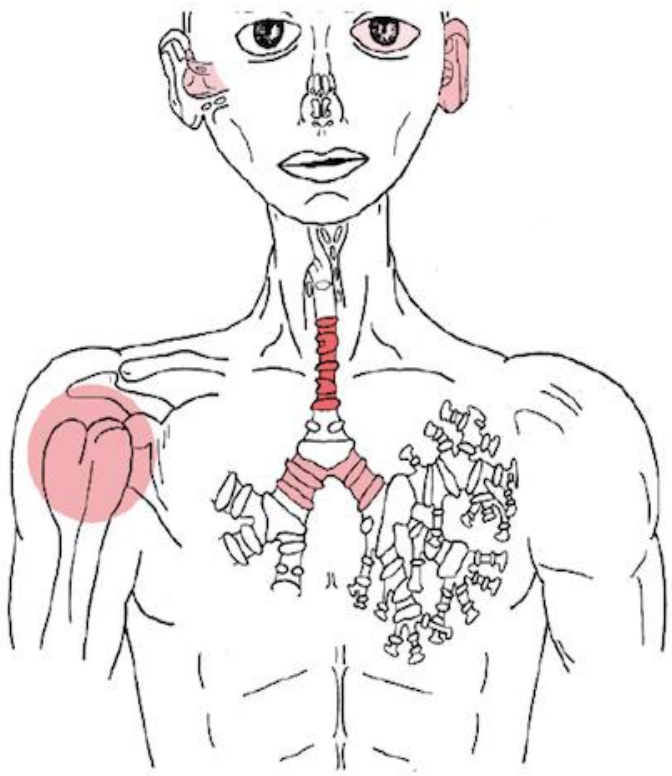
Most Common Type of RP

Not!!

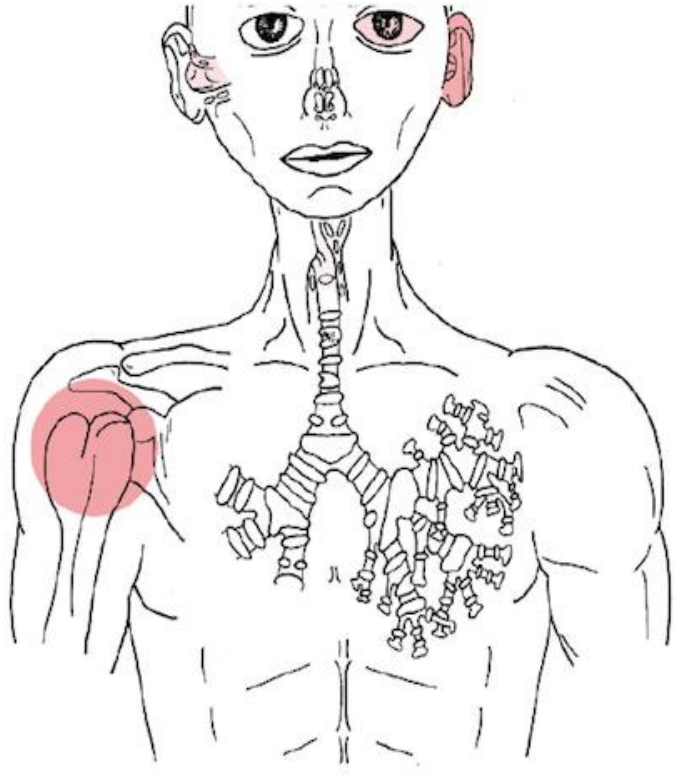
RP Can be Divided in Three Clinical Phenotypes



Type I

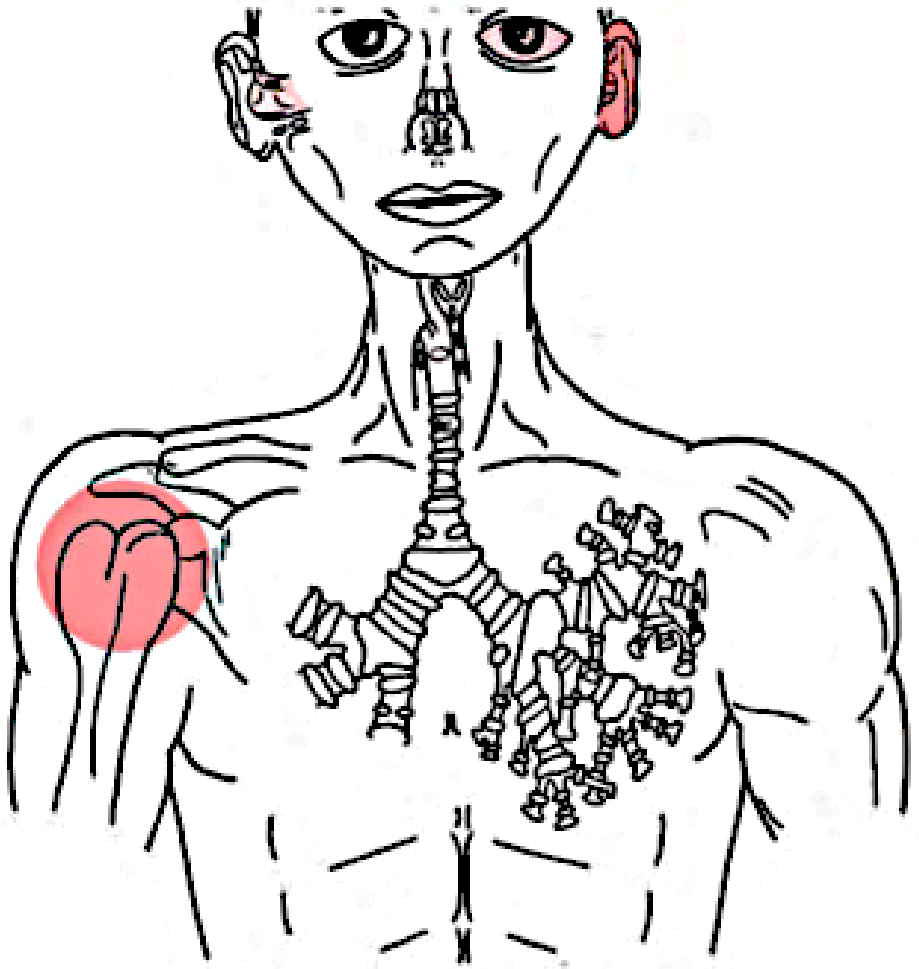


Type II

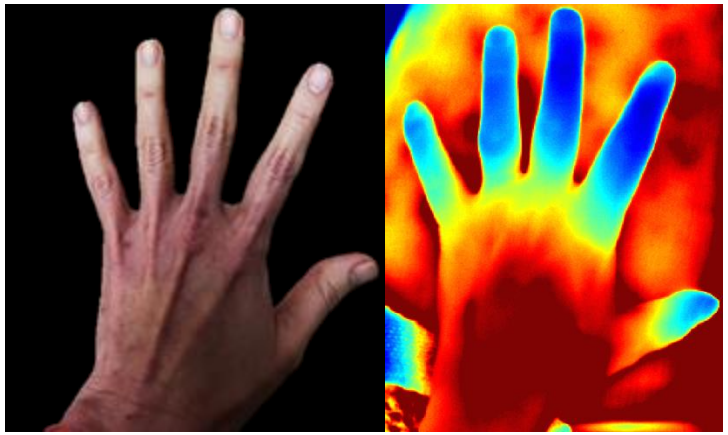


Type III

Ferrada et al, Arthritis Rheumatol. 2020 Aug;72(8):1396-1402



- Arthalgias without arthritis
- Raynaud's
- Dry eyes/Dry mouth
- Myalgias
- Muscle weakness
- Opthalmoplegia
- GI manifestations
- **MAGIC**
- **VEXAS**



Ferrada et al, Arthritis Rheumatol. 2020 Aug;72(8):1396-1402

Dont Forget to Ask About Genital Ulcers!!!

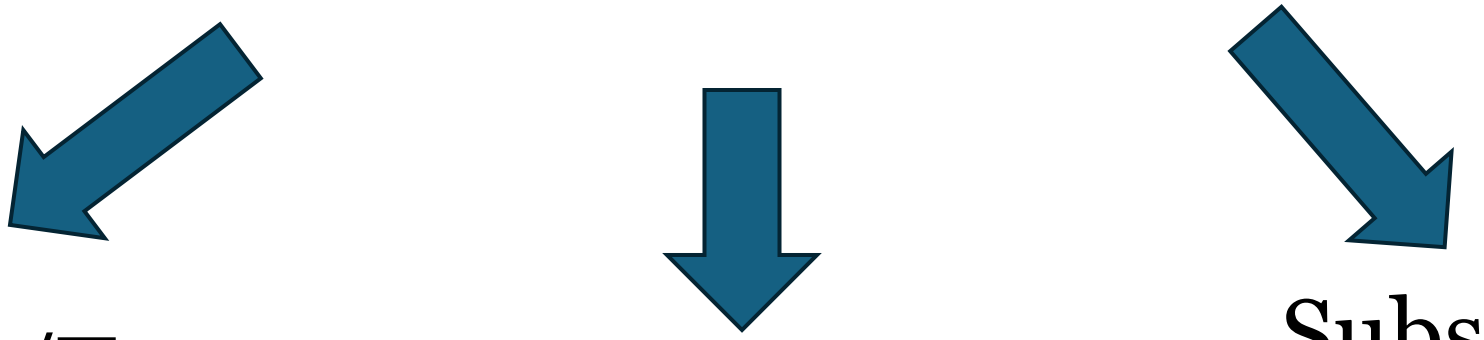
MAGIC

Clinical Evaluation

Review of systems

Pictures

Physical Exam



Imaging/Tests

Rule Out
Other Diagnoses

Subspecialities
Evaluation

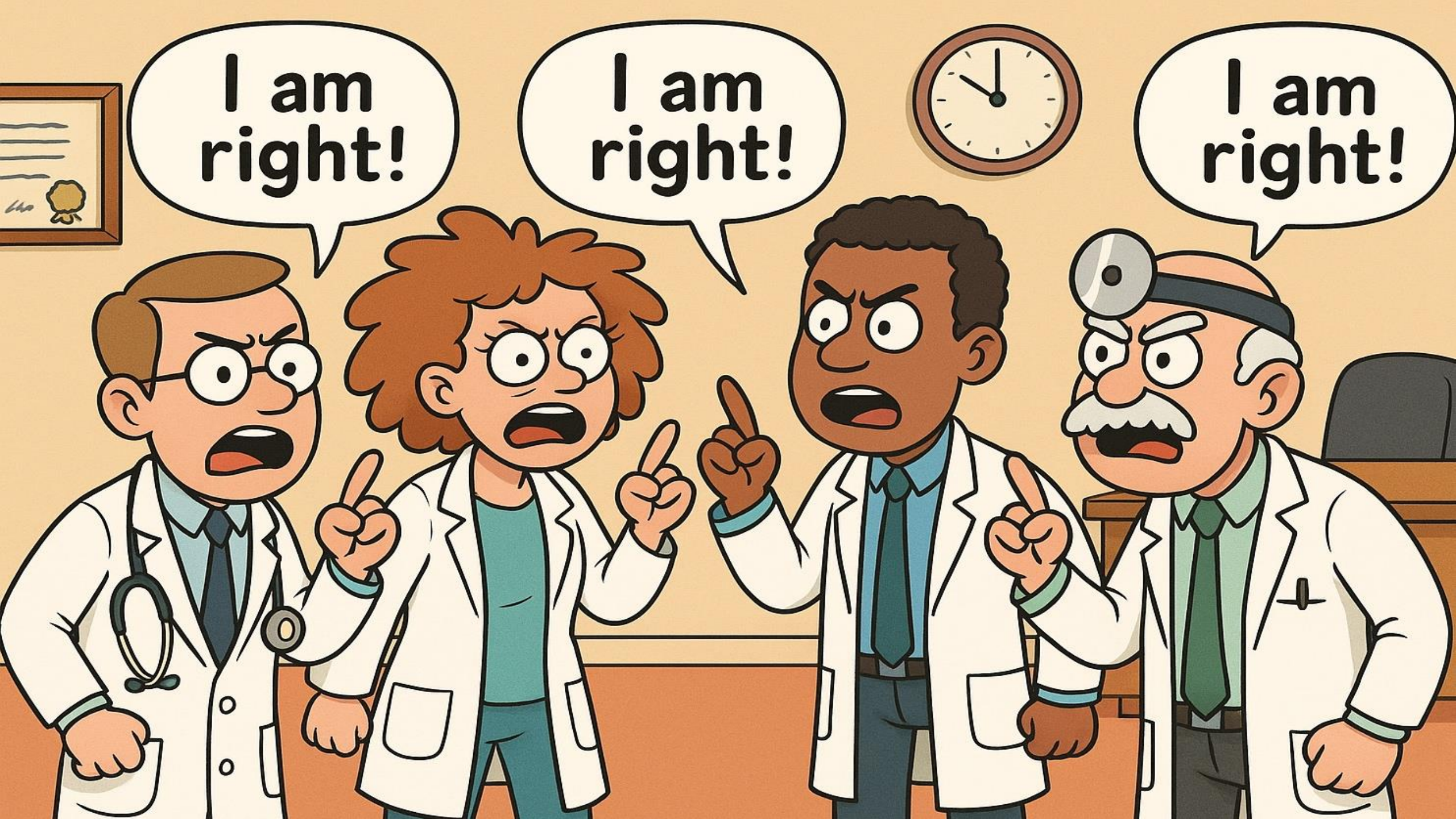
Recommended Care for Patients with RP

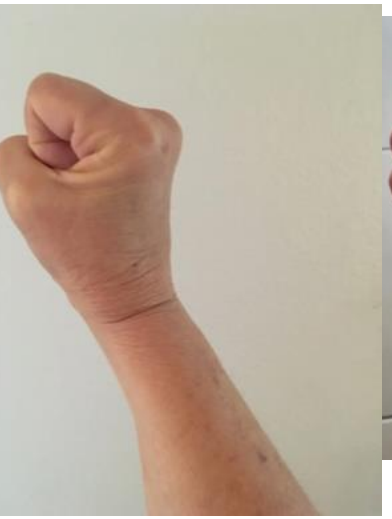
- **Audiology evaluation**
  - Inner ear involvement
- **Echocardiogram**
  - Mitral and aortic valve involvement
- **Ophthalmology evaluation**
  - Episcleritis, retinal detachment
- **Dynamic CT scan**
  - TM, BM, tracheal and bronchial thickening or stenosis, air trapping
- **ENT**
  - Upper airway disease

- **Most common sDMARD's**
  - **MTX, Leflunomide and MMF**
- **Most common bDMARD's**
  - **TNF and anti-IL6**

Future Research

Clinical Phenotype

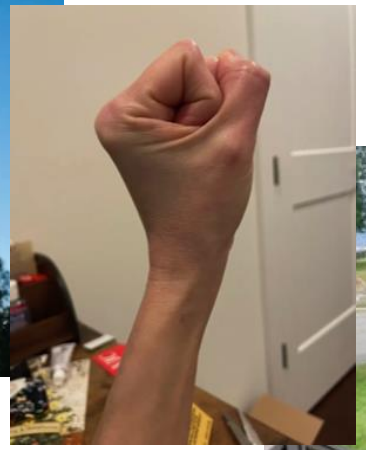

I am right!
I am right!
I am right!



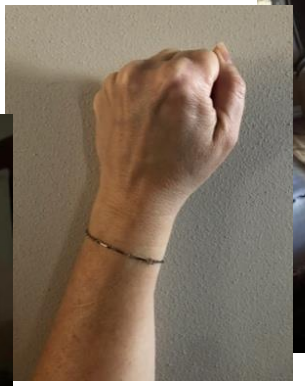




The Belief That the Future will be Better Because We Have the Power to Make it Happen


HOPE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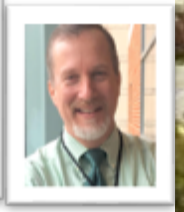

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