RELAPSING POLYCHONDRITIS

By (in order of presentation):

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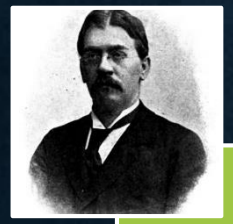
OUTLINE

- Introduction
- History and Epidemiology
- Pathogenesis
- Clinical manifestations
- Diagnosis and Prognosis
- Treatments
- Conclusions

INTRODUCTION

- Definition
  - RP is an immune-mediated systemic disease characterized by recurrent inflammatory episodes of cartilaginous and proteoglycan-rich tissues
- Onset
  - acute painful inflammatory crisis
  - spontaneous remission of variable duration
- Associations with
  - Autoimmune disorders
  - Rheumatoid arthritis

HISTORY



Jaksch Wartenhorst

- 1923
- Jaksch Wartenhorst
- Polychondropathia

- 1960
- Pearson et al.
- Relapsing polychondritis

Lawrence McAdam

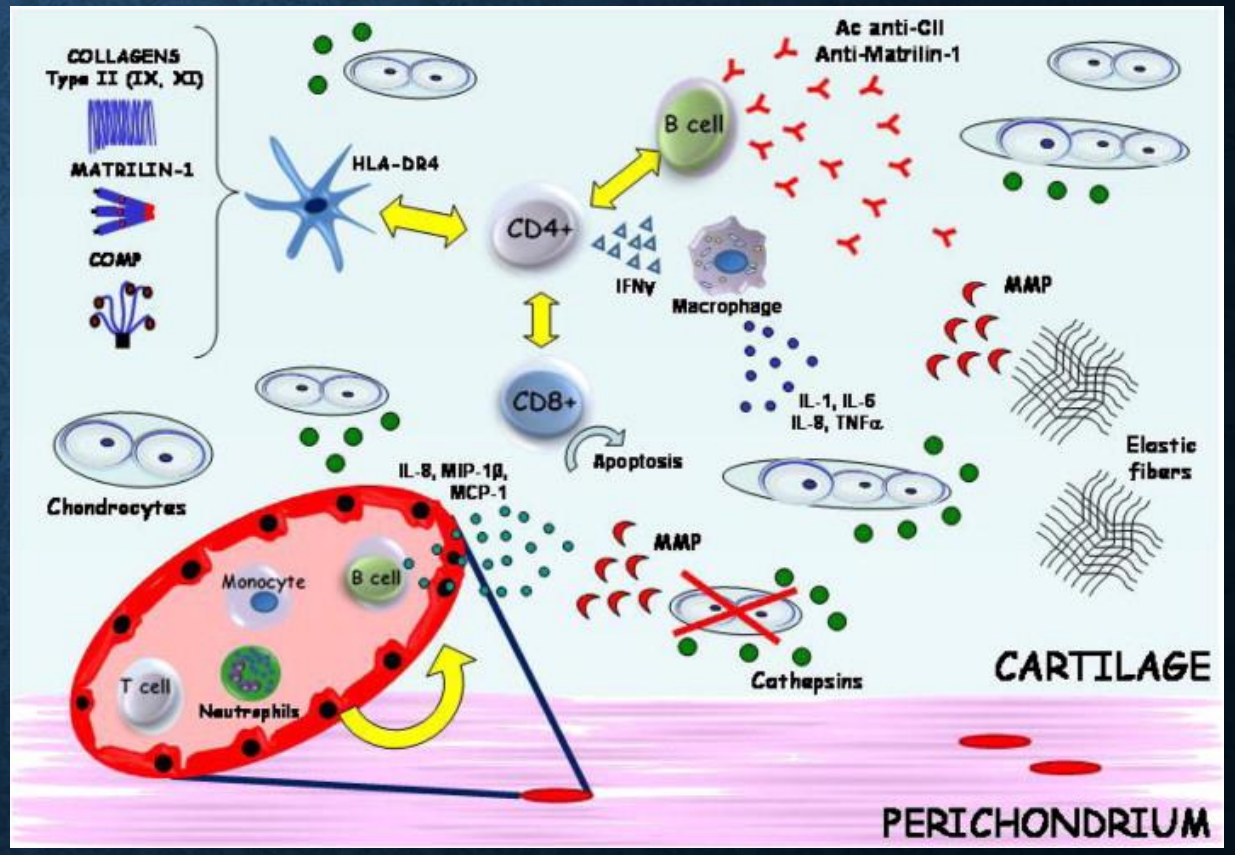
- 1976
- McAdam et al.
- The first diagnostic criteria for RP

EPIDEMIOLOGY

- Incidence
- Median age of onset
- Pediatric RP
  - Frequency
  - Age of onset
- Pregnancy and RP
- Frequency among genders
- Frequency among ethnic groups

PATHOGENESIS

- Major risk allele
  - HLA-DR4
- No familial transmission
- Humoral and cell-mediated immune systems
- Autoimmunity



Clinical Manifestations

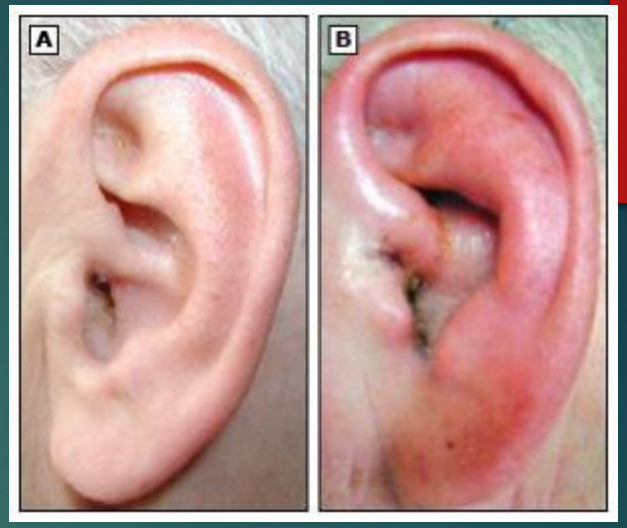
PRESENT BY:

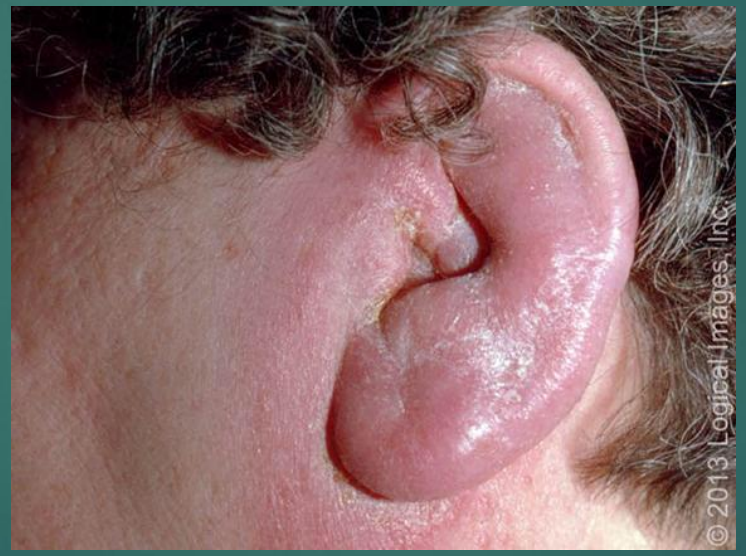
HAMED GHORANI

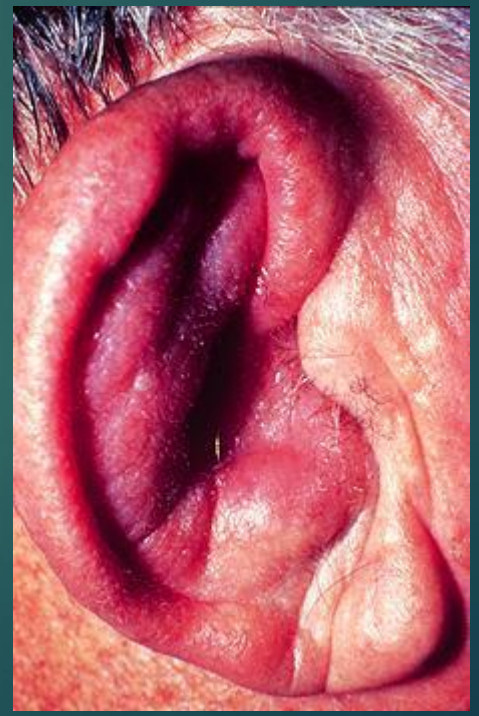
1. Chondritis: Ear, Nose, Large Airways
2. Arthropathy
3. Ocular Involvement
4. Neurologic Manifestations
5. Renal Manifestations
6. Dermatological Manifestations
7. Cardiovascular Manifestations
8. Gastrointestinal tract Involvement

Ear involvement

- MONO- OR BILATERAL AURICULAR CHONDRITIS
- THE MOST COMMON FEATURE
- CAULIFLOWER EAR
- HEARING LOSS
  - ✓ Conductive
  - ✓ Sensorineural
- OTITIS EXTERNA
- CHRONIC MYRINGITIS
- TINNITUS

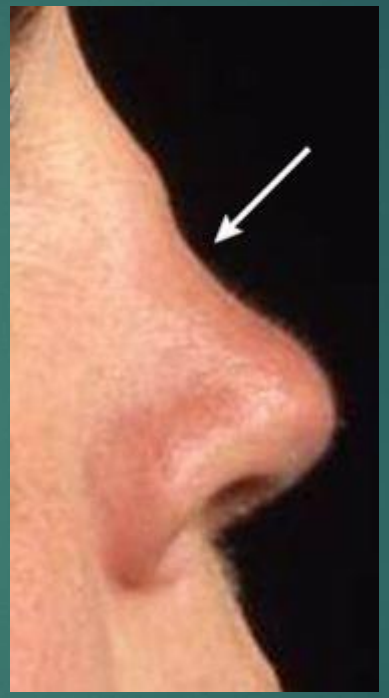






Nasal Involvement

- STUFFINESS
- CRUSTING
- RHINORRHEA
- EPISTAXIS
- HYPOGEUSIA
- SADDLE NOSE DEFORMITY
  - Female
  - age<50



Laryngotracheobronchial involvement

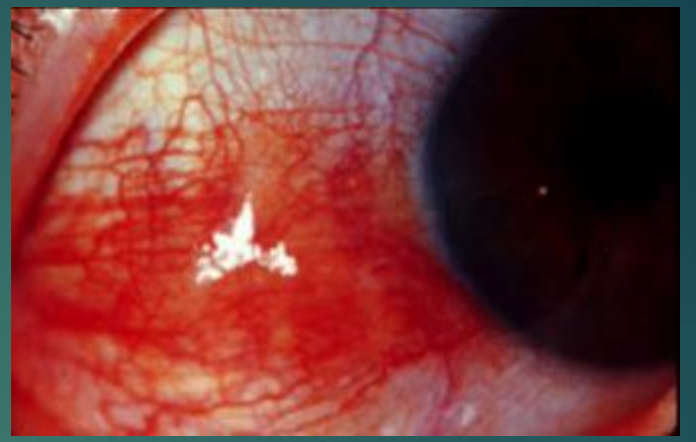
- LARYNGOMALACIA
  - Hoarseness of voice
  - Non-productive cough
  - Stridor
  - Wheezing
- TRACHEOBRONCHOMALACIA
  - **Major cause of mortality and morbidity**
  - Secondary pulmonary infection
- NECROTIZING SIALOMETAPLASIA OF LARYNX
- OSA
- DYNAMIC COLLAPSE OF LARYNX & TRACHEA

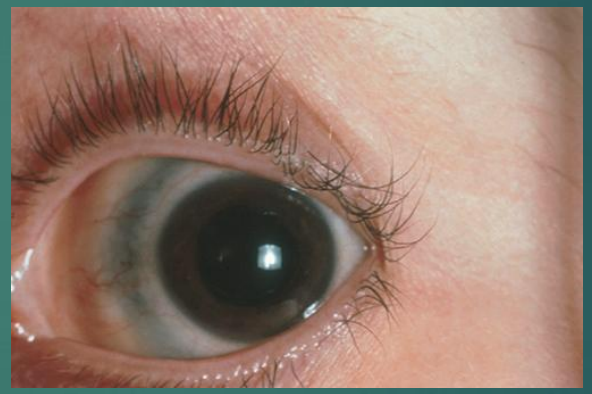
Arthropathy

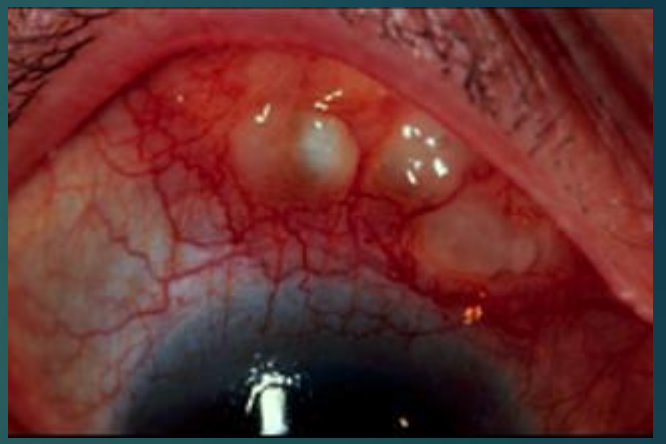
- ACUTE ASYMMETRIC INTERMITTENT OLIGO- OR POLYARTHRITIS
  - MCP
  - PIP
  - Knee
  - Parasternal joints: Sternoclavicular, Costochondral, Manubriosternal
- NON-EROSIVE ARTHRITIS
- NON-INFLAMMATORY SYNOVIAL FLUID ASPIRATES

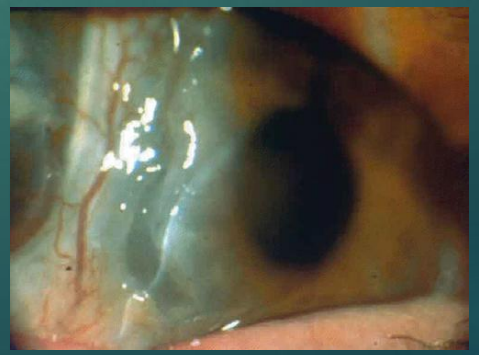
Ocular Manifestations

- EPISCLERITIS
- SCLERITIS
  - Diffuse anterior scleritis
  - Necrotizing anterior or posterior scleritis
- SCLEROMALACIA
- PERIPHERAL ULCERATIVE KERATITIS
- UVEITIS
- SALMON PATCH
  - Benign reactive lymphoid hyperplasia in conjunctiva
- PROPTOSIS
- OPTIC NEURITIS
- CATARACT







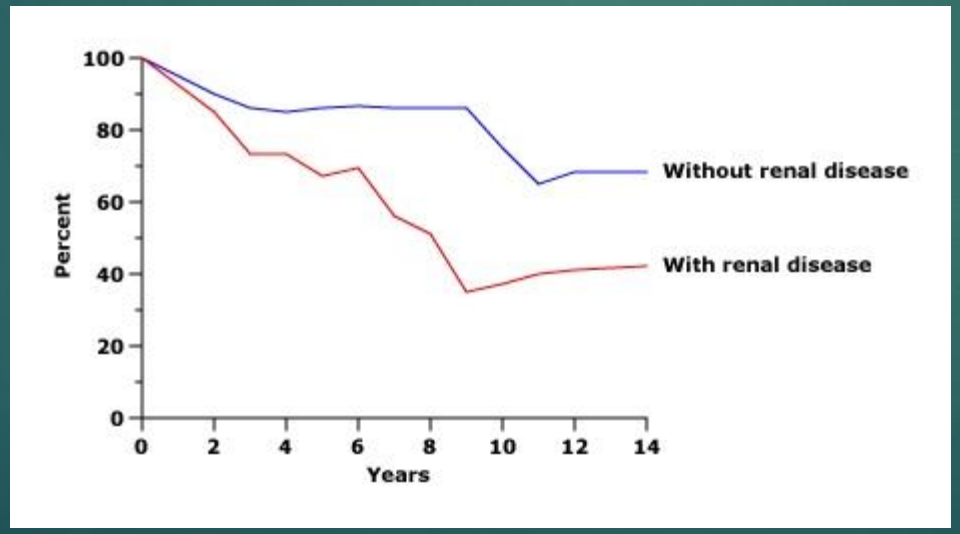


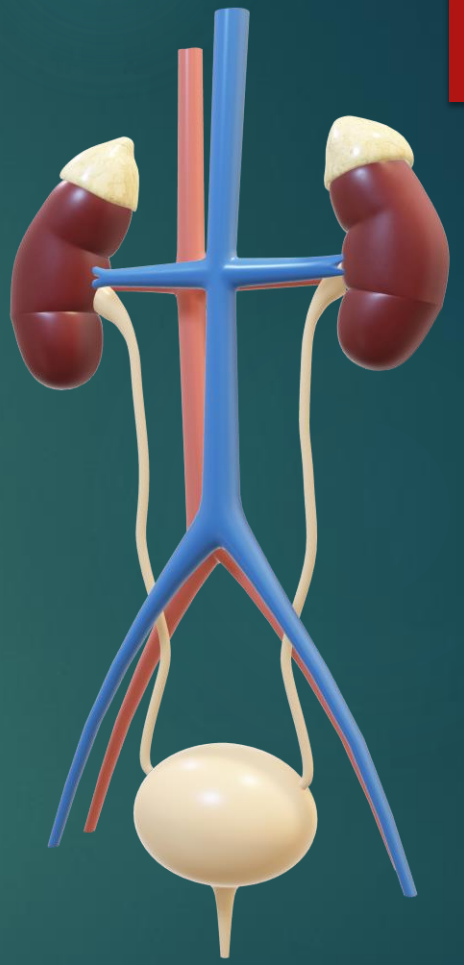
Neurologic Manifestations

- CRANIAL NEUROPATHIES: CN V & CN VII
- VASCULITIS
- HEADACHE
- ASEPTIC MENINGITIS
- LIMBIC ENCEPHALITIS
- HEMIPLEGIA
- ATAXIA
- SEIZURE
- COGNITIVE DYSFUNCTION
  - ✓ Fulminant, multisystem presentation, subacute course, CNS vasculitis
  - ✓ Insidious course without associated constitutional or systemic symptoms

Renal Manifestations

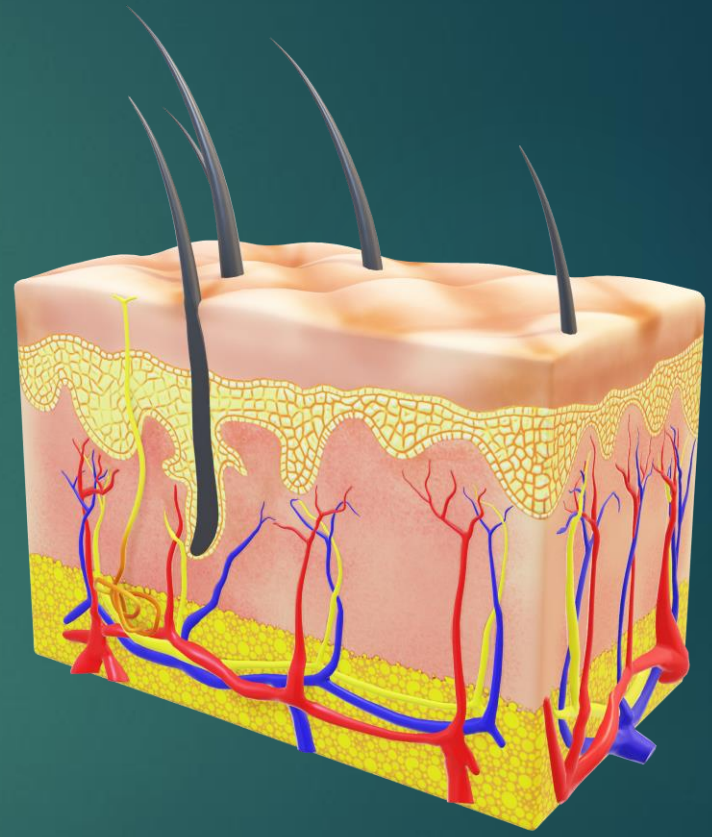
- POOR PROGNOSIS
- MESANGIAL EXPANSION
- IGA NEPHROPATHY
- TUBULOINTERSTITIAL NEPHRITIS
- SEGMENTAL NECROTIZING CRESCENTIC GN.
- MEMBRANOUS NEPHROPATHY





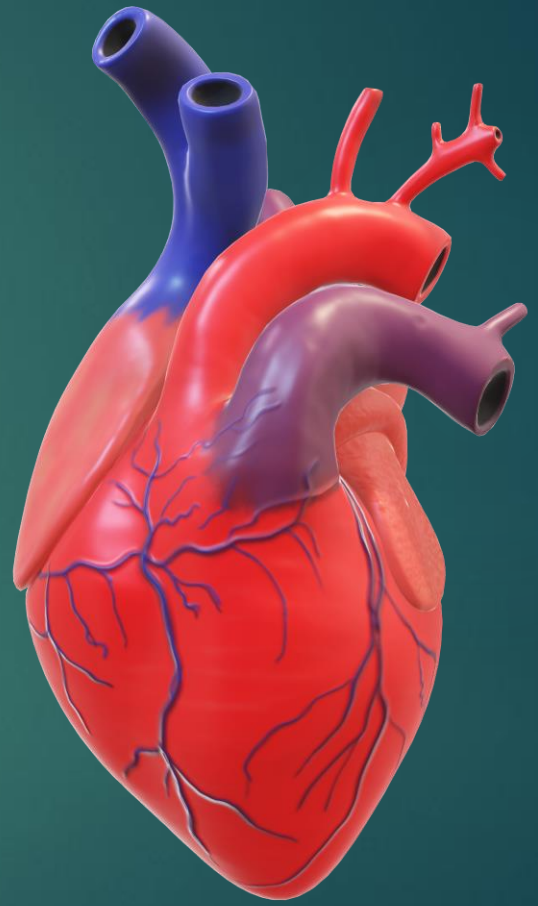
Dermatologic Manifestations

- APHTOSIS
- NODULES IN THE LIMBS
- PURPURA AND PAPULES
- LIVEDO RETICULARIS
- DISTAL ULCERATIONS AND NECROSIS
- TENSE URTICARIAL PAPULES
- **MAGIC**
  - Relapsing polychondritis
  - Behcet's disease
- MDS
  - Sweet syndrome (neutrophilic dermatosis)



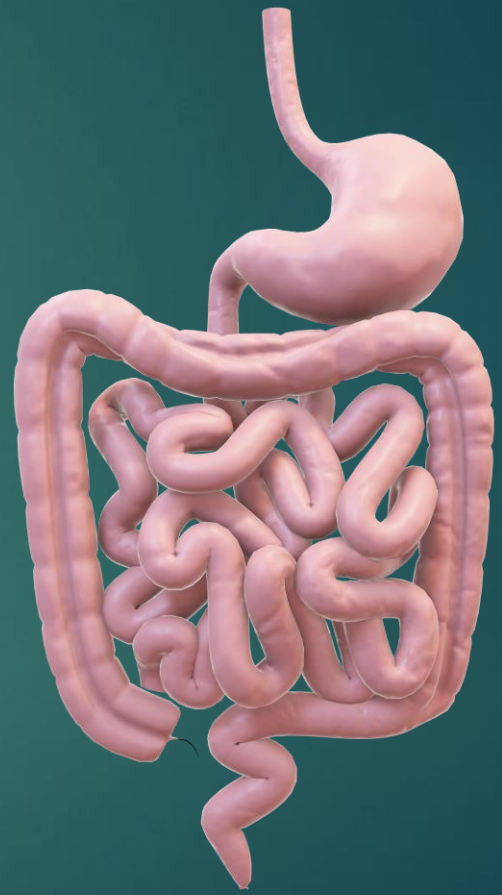
Cardiovascular Manifestations

- VALVULAR HEART DISEASE
  - AI : 4-6%
  - MR : 2-4%
- AORTIC ANEURYSM
- AORTIC DISSECTION
- MYOCARDITIS
- PERICARDITIS
- AV BLOCK
- SYSTEMIC VASCULITIS
  - Cutaneous leukocytoclastic vasculitis
  - Large vessel vasculitis



Gastrointestinal tract involvement

- DUE TO SYSTEMIC VASCULITIS
- IBD
  - Crohn's disease
  - UC
- SYSTEMIC SCLEROSIS
- DIABETIC AUTONOMIC DYSFUNCTION
- DYSPHAGIA
- PSC
- PBC
- PNEUMATOSIS CYSTOIDES



Associated disorders

- AUTOIMMUNE DISORDERS
  - SLE
  - SS
  - MCTD
  - Sjogren's syndrome
  - Dermatomyositis
- RHEUMATOLOGICAL DISEASES
  - RA
  - Spondyloarthropathy
  - Vasculitis
- MDS
- SOLID TUMORS: BLADDER, BREAST, LUNG COLON, PANCREAS

Diagnosis and Prognosis

The diagnosis of RP is a real challenge for clinicians, because of the pleomorphic nature and insidious onset of the disease

Authors, Year and Reference	Suggested Criteria
Mc Adam et al. 1976 [3]	At least three clinical features among auricular chondritis, nonerosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, audiovestibular damage; histologic confirmation not required
Damiani and Levine 1979 [4]	At least one of the six clinical features suggested by Mc Adam et al. [3] plus histological confirmation *or* two of the six clinical features suggested by Mc Adam et al. [3] plus positive response to administration of corticosteroids or dapsone
Michet et al. 1986 [5]	Confirmed inflammation in two of three cartilages among auricular, nasal or laryngotracheal *or* proven inflammation in one of the above cartilages plus two other minor criteria among hearing loss, ocular inflammation, vestibulary disfunction, seronegative arthritis

Laboratory findings

- C-reactive protein and erythrocyte sedimentation rate
- ANA,RF,Antiph
- ANCA
- anti-type II collagen antibodies
- anti-matrilin-1 antibodies
- COMP levels
- X-ray,CT,MRI
- Bronchoscopy
- UA,Cr
- Echocardiography
- Bone scintigraphy

Relapsing Polychondritis Disease Activity Index(RPDAI):

- The RPDAI score is made up of 27 items with individual weights ranging from 1 to 24 and a maximum theoretical RPDAI score of 265, taking into account disease manifestations in a 28-day period (online scoring at www.RPDAI.org)

Survival rates:

- 70% after five years
- 94% after eight years
- 91% after 10 years

TREATMENT OF RP

OPTIONS:

- Pharmacologic therapy
- Surgical appraoch

PHARMACOLOGIC THERAPY

- Including:

1. NSAID
2. SYSTEMIC CS
3. METHOTREXATE
4. CYCLOSPORINE
5. CYCLOPHOSPHAMIDE
6. AZATHIOPRINE
7. BIOLOGICS or second line option:(ANTI-TNF/TOCILIZOMAB/ABATACEPT/RITUXIMAB/ANAKINRA)
8. OTHERS: (6-MERCAPTOPURINE/PLASMAPHRESIS/ANTI CD4/MINOXIDINE/PENICILLAMINE/IVIG/LEFUNOMIDE)

1. In non severe RP + control of pain and inflammation: NSAID

2. Mild manifestation(only nose,ext.ears,joint):

- dapsone(50-100 in max dose 200 mg once daily)
- colchicine(.06 mg 2-4 times in day)

3. NSAID resistant or severe RP (ocular,laryngotracheal,cardiac involvement ,sever vasculitis,): systemic CS (oral : perdenisone 0.25-1 mg/kg/d) (methylperdnisolone 500-1000 mg/d)

4. Second line option: (CS intolerant,lack of response to CS,need for CS sparing therapy) : anti TNF-rituximab-anakinra-tocilizumab-abatacept)

SURGICAL APPROACH

- Severe bronchial stenosis (tracheostomy or stent)
- Intractable cardiac failure because of valve regurgitation or aortic aneurism