

Many Faces of Rheumatic Disorders

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Agenda and Objectives

- A few words about Pierre-Auguste Renoir
- What do we know about arthritis and autoimmune diseases
- Major types of autoimmune connective tissue diseases
 - Rheumatoid Arthritis
 - Lupus
 - Scleroderma
 - Sjogren's syndrome
 - Polymyositis and Dermatomyositis
 - Vasculitis
- Snapshot of clinical features and management

Pierre-Auguste Renoir (1841-1919)



- *"For me a picture has to be something pleasant, delightful, and pretty - yes, pretty. There are enough unpleasant things in the world without us producing even more." ~*



- *"One must from time to time attempt things that are beyond one's capacity." ~ Pierre-Auguste Renoir*

Struggles through Pain





Renoir's *Luncheon of the Boating Party*, 1881



The Umbrellas-1881



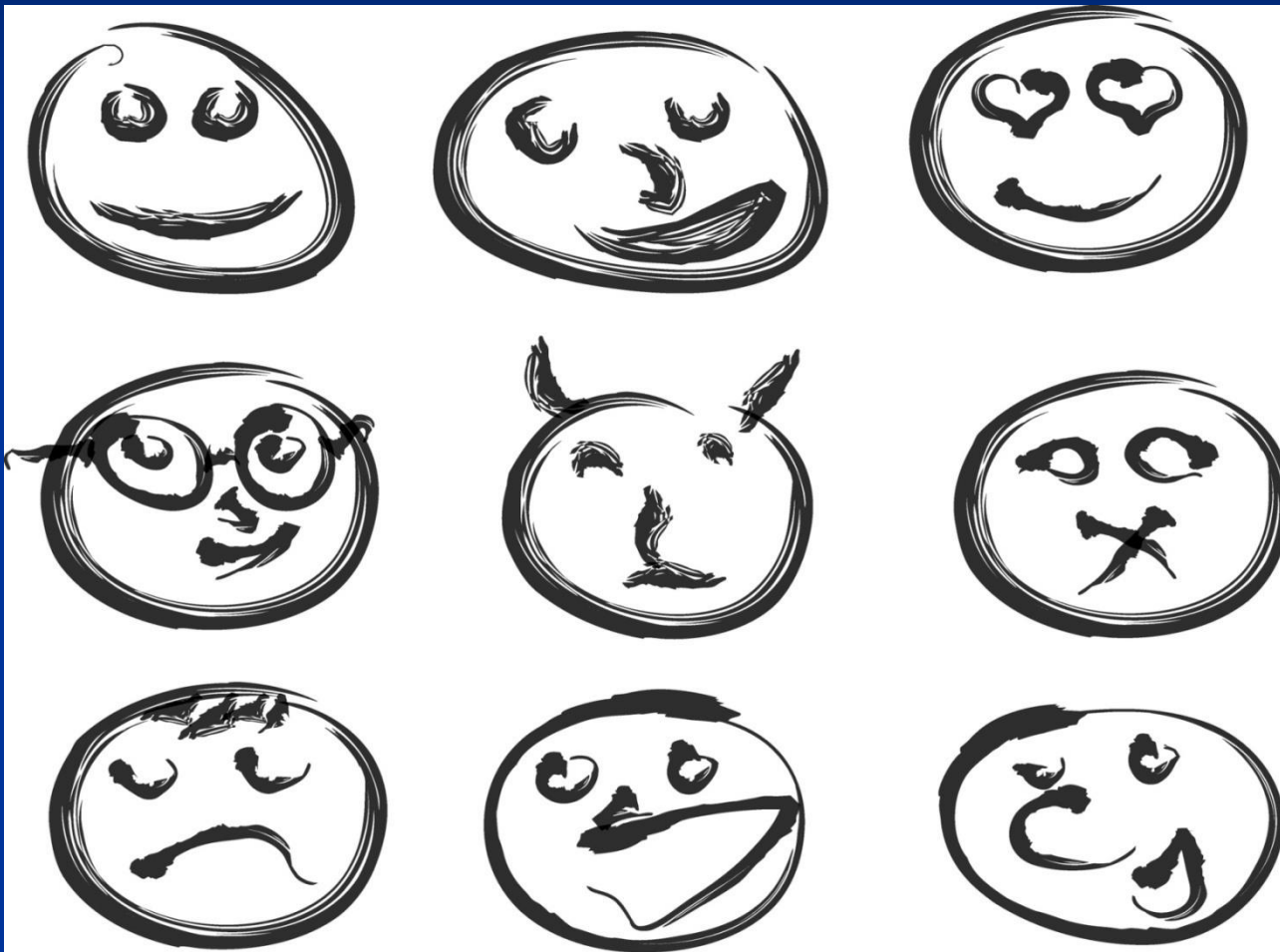
“The pain passes, but the beauty remains.”

~ Pierre-Auguste Renoir

It can affect all of us



It is not pleasant!

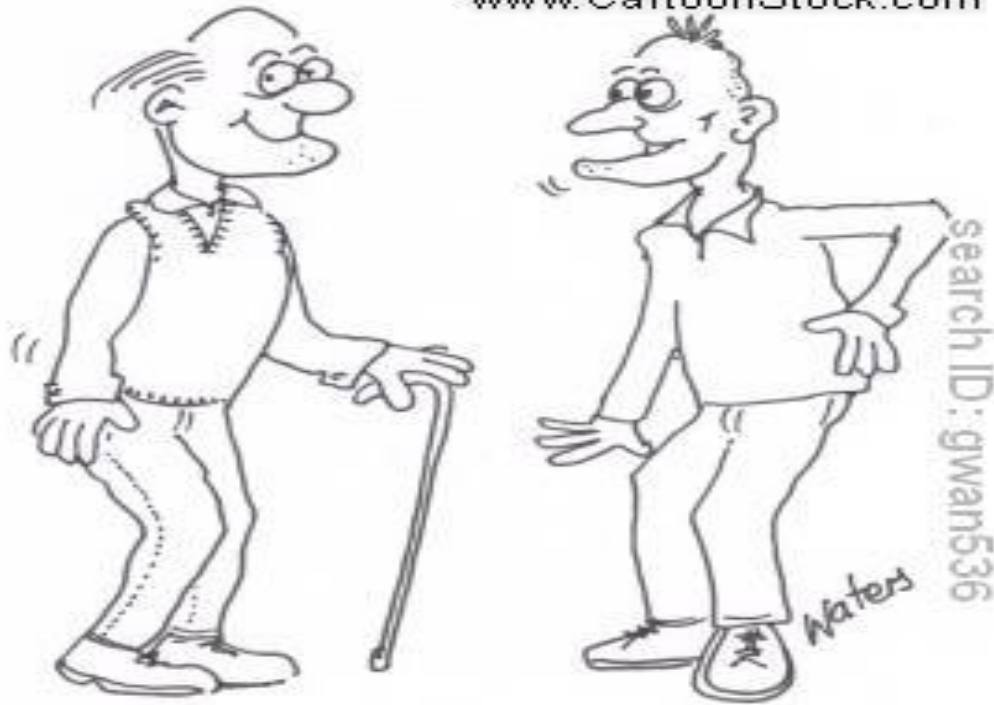


Arthritis does not spare anyone



Arthritis hurts!

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"When you get older everything hurts...
and what doesn't hurt doesn't work!"

Arthritis

- Arthritis is a disorder which can cause mild to severe pain in the joints, joint tenderness and swelling
- Approximately 50 million Americans have some type of arthritis or related condition
- Huge socioeconomic impact on society resulting in costs of \$128 billion annually, and continues to be the most common cause of disability

22% Of US Adults Have Arthritis

Rheumatology
Morning Wire

Prepared exclusively for
members of
 AMERICAN COLLEGE
OF RHEUMATOLOGY
EDUCATION • TREATMENT • RESEARCH

In affiliation with
 CUSTOM
BRIEFINGS

Today's News for the American College of Rheumatology from Newspapers, TV, Radio and Journals

- 50 million people with the joint diseases

Centers for Disease Control and Prevention

MMWR

Morbidity and Mortality Weekly Report

Weekly / Vol. 59 / No. 39

October 8, 2010

**Prevalence of Doctor-Diagnosed Arthritis and Arthritis-Attributable Activity
Limitation — United States, 2007–2009**

- Prevalence of doctor-diagnosed arthritis and arthritis-attributable activity limitation (AAAL) is expected to increase significantly by 2030

Who to blame?

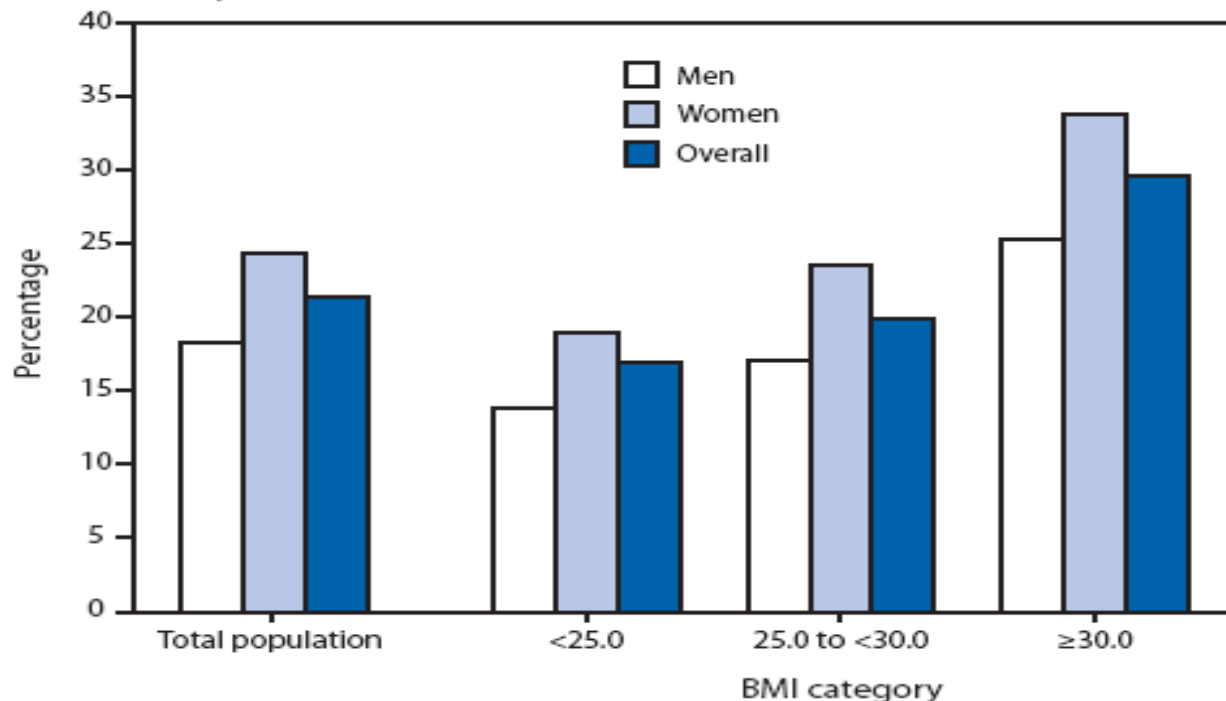
- The driving factor for the rise in arthritis patients appears to be obesity
- Among obese, one in three women and one in four men have arthritis, roughly double the proportion among those whose weight is normal
- " What's more, "obesity particularly plays a role in the onset of knee osteoarthritis, the most common form of the disorder."

National Health Interview Survey for 2007-2009

- Among major findings from the
 - 22.2% of adults aged 18 and older say doctors have diagnosed them with arthritis, 49.9 million people
 - 42.4% of people with diagnosed arthritis, or 21.1 million people suffer limitations on physical activity because of their disease
 - Among the obese, 33.8% of women and 25.2% of men , rates were close to double that of people who are underweight or normal weight, 13.8% for men and 18.9% for women

Findings of Arthritis Survey

FIGURE. Age-adjusted* prevalence of doctor-diagnosed arthritis among adults, by sex and body mass index (BMI) category† — National Health Interview Survey, United States, 2007–2009



* Age adjusted to the 2000 U.S. projected adult population, using three age groups: 18–44 years, 45–64 years, and ≥65 years.

† BMI = weight (kg) / height (m²). Categorized as follows: underweight/normal weight (<25.0), overweight (25.0 to <30.0), and obese (≥30.0).

Who is most affected ?

- Focusing on specifics, the investigators noted that "after adjustment for age, the prevalence of arthritis-related activity limitations were highest for
 - Body mass index (BMI) was above 40 (52.9%)
 - With low education levels (52%)
 - Inactive (51.2%)
 - Smokers (47.6%)
 - Non-Hispanic blacks (45.5%)

What's being predicated ?

- "'With the aging population and continued high prevalence of obesity,' arthritis is predicted to increase significantly over the next 20 years
- Projecting are that "the number of adults with arthritis will hit 67 million by 2030"
- Many of these people will "experience arthritis-related limitations in activity"

Autoimmune arthritis

- Disorder of immune system
- Rheumatoid Arthritis is the classic example
- Destruction of joint is fast and aggressive, MRI and ultrasound can detect damage in few months
- Systemic features-fever, fatigue, weight loss
- Unaffected by inactivity, improves with movement and exercises
- It can affect internal organs

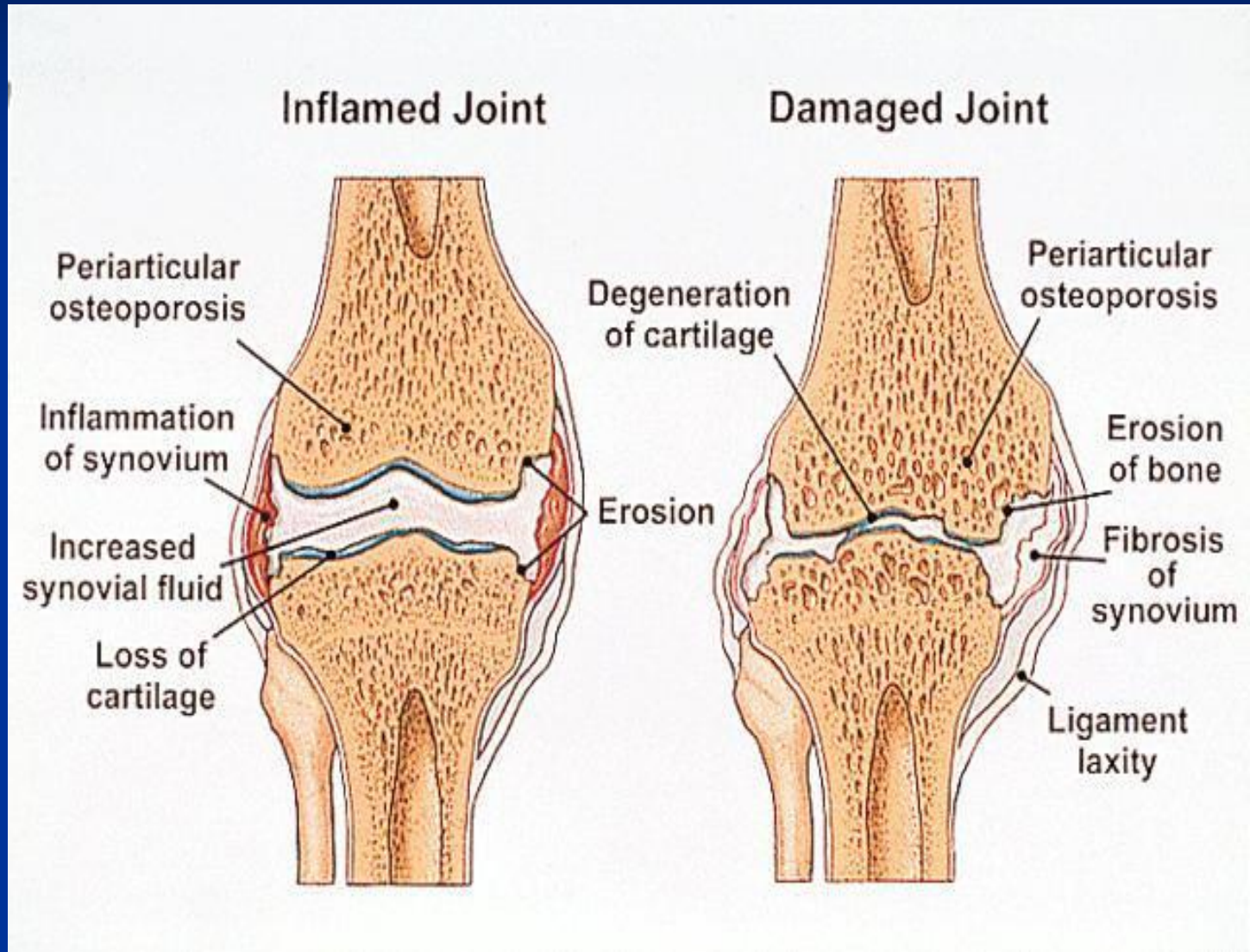
Cause of Autoimmune arthritis

- Exact cause – Unknown
- Environmental factors triggers the susceptible individual who carry certain genes
- Risk factors
 - female gender
 - Poor dental hygiene
 - Genes
- Triggers include
 - Smoking
 - Viral infection
 - Post partum period
 - Stress

Rheumatoid arthritis

- Classic example of chronic type of inflammatory arthritis
 - Onset may be abrupt or insidious
 - Prolonged morning stiffness (>1 hr)
 - Polyarthritits, >5 swollen and tender joints
 - Duration of persistent symptoms for >8 weeks
 - Additive and progressive pattern
 - Symmetrical or bilateral involvement
- Chronic inflammation can lead to deterioration, pain and limited movement
- Approximately affecting 0.5% of population with women outnumbering men, ratio 3:1, mean age 48 years

Joint destruction



Rheumatoid arthritis in a young female



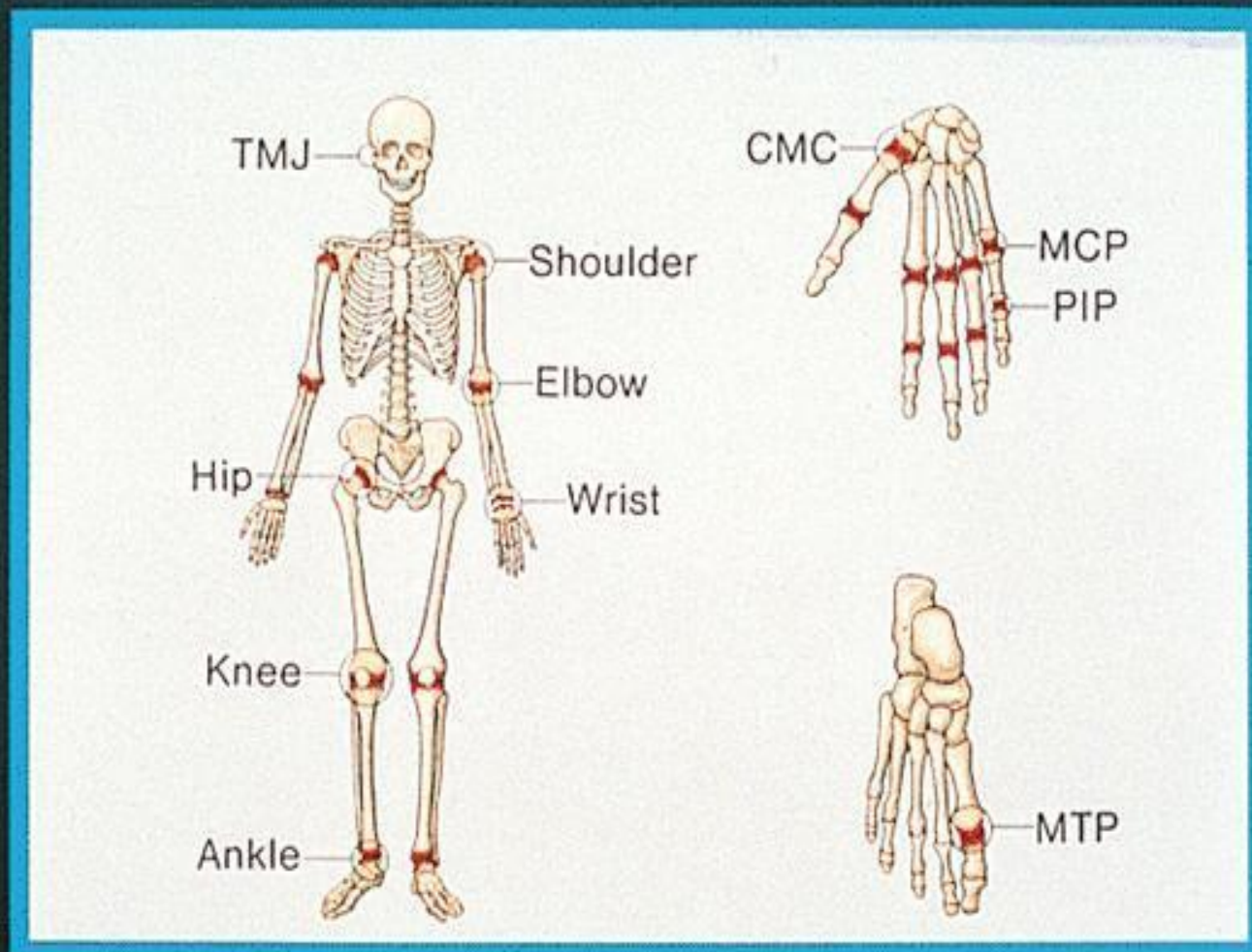
Laboratory findings in Rheumatoid Arthritis

- Elevated acute phase reactants
 - High ESR,CRP
 - Serum amyloid
 - Low hemoglobin
 - Increased platelet count
 - Increased alkaline phosphatase
 - Low Wbc or pancytopenia with complications
- Serologic markers
 - Rheumatoid factor + in >70%
 - ANA +50%-70%
 - Anti CCP (highly specific->85%)

Radiologic tests and Treatment

- Radiology
 - Early synovitis detected with Ultrasound and MRI
 - Erosions are punched out lesions on bony surfaces
 - X-rays of hands and feet as baseline
- Survey of Cervical spine for instability

Arthritis has distinct pattern and



Pattern of joint involvement

Arthritis



OA



RA



PsA



CPPD

Management of Rheumatoid Arthritis

- Early Referral and Treatment
 - Bridging therapy: Nsaids and prednisone
 - DMARDs: Plaquenil, Sulfasalazine, Methotrexate
- Revolutionary biologic therapy
 - Etanercept, Infliximab, Adalimumab etc
 - Rituximab, Abatacept and Tocilizumab etc

Other faces of autoimmune arthritis

- Lupus
- Scleroderma
- Sjogren's syndrome
- Polymyositis and Dermatomyositis
- Vasculitis

Lupus

(systemic lupus erythematosus)

- Systemic autoimmune disease with a wide variety of clinical features
- Considerable variation- mild(most) to life threatening (rare)
- Nine times more common in women
- Systemic disease- can affect joints, skin, kidneys, lungs, brain
- 240,000 Americans -- 90 percent of whom are females
- Mainly affects women of childbearing age
- More in African American, Asian and Latino populations than Whites

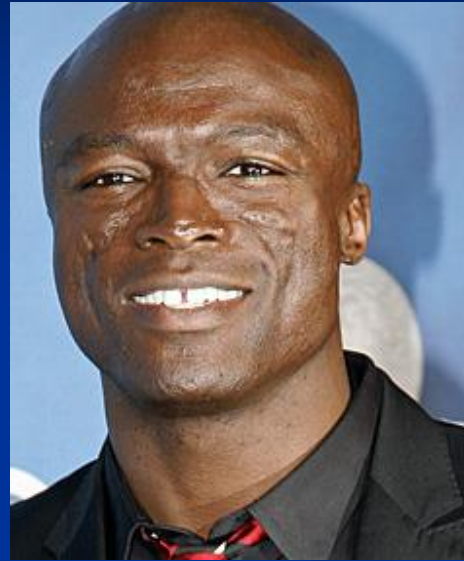
Clinical Features of Lupus

- Fatigue, hair loss, dry mouth and eyes
- Muscle and joint pain, muscle weakness
- Fever
- Oral and nasal ulcers
- Raynauds
- Kidney failure and protein loss in urine
- Chest pain from inflammation of lung lining
- Seizures, Headache
- Decreased blood cell and
- Stroke and venous blood clots

Laboratory findings in Lupus

- Suggestive of inflammation
 - Low HB, Low WBC, Low Platelets
 - ESR- high, but CRP is normal
 - Prolonged APTT
 - Creatinine and elevated protein in urine
- Serologic tests
 - Positive ANA (95%), Anti DsDNA (50-70%)
 - Low complements
- Biopsy: Renal and lung, Skin

Discoid Lupus

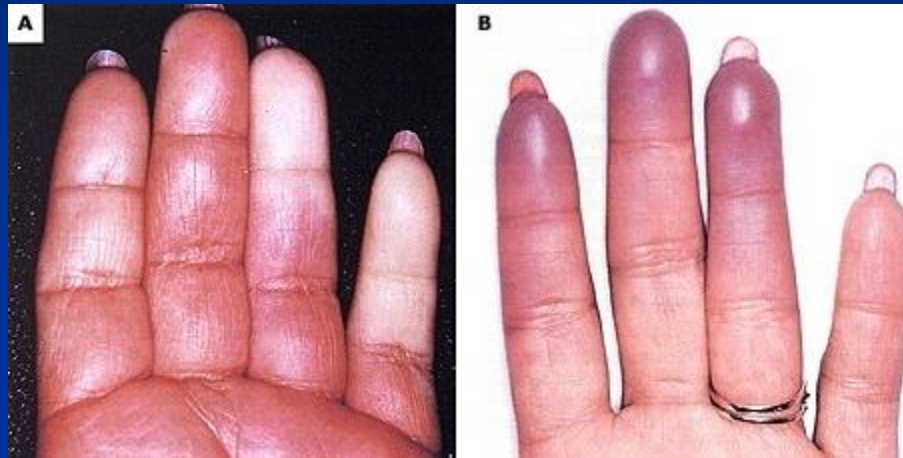


Lupus Skin changes

Malar (Butterfly) rash and oral lesions



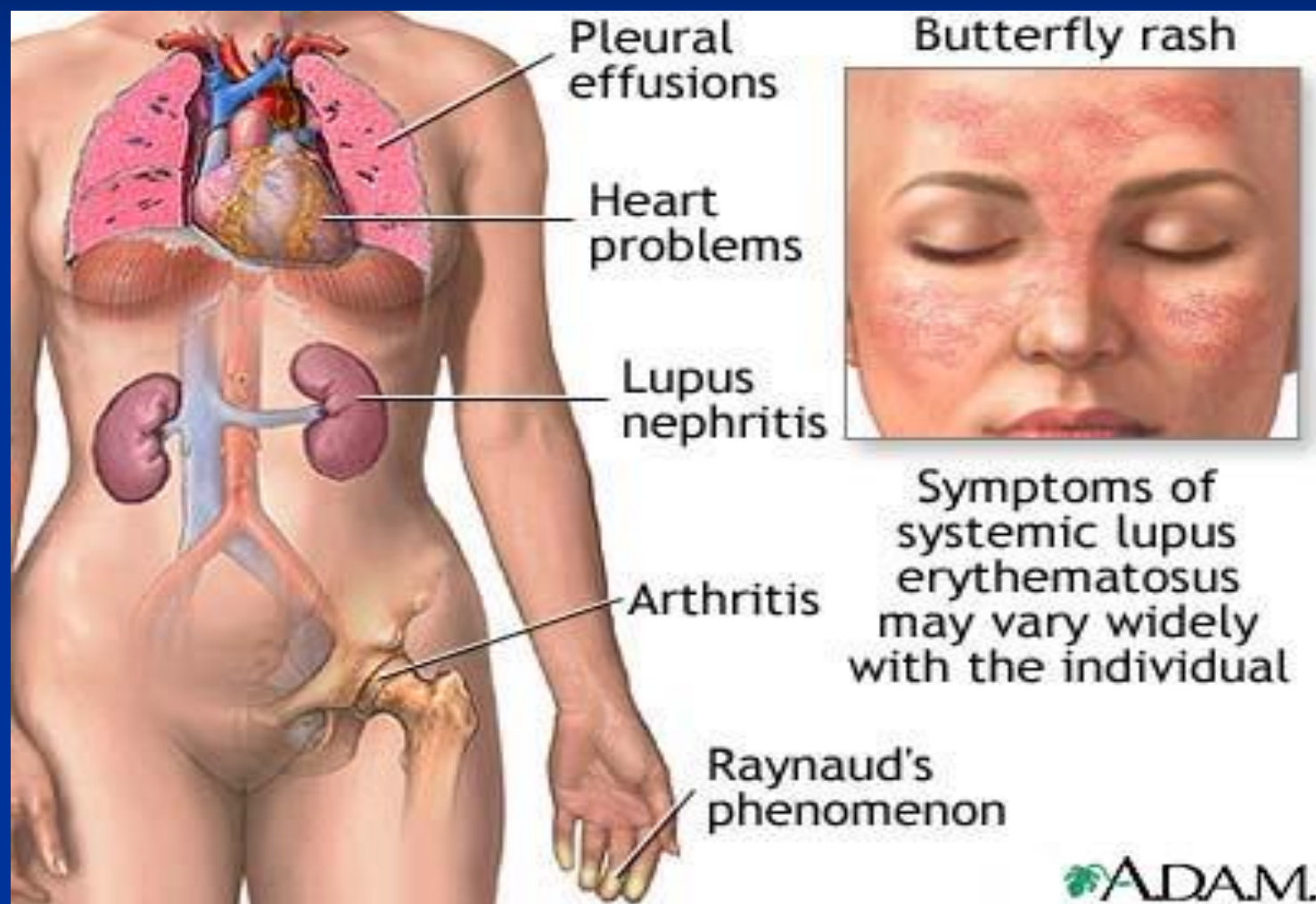
Rash on hands and Raynauds



Lupus can also affect men and arthritis can be deforming

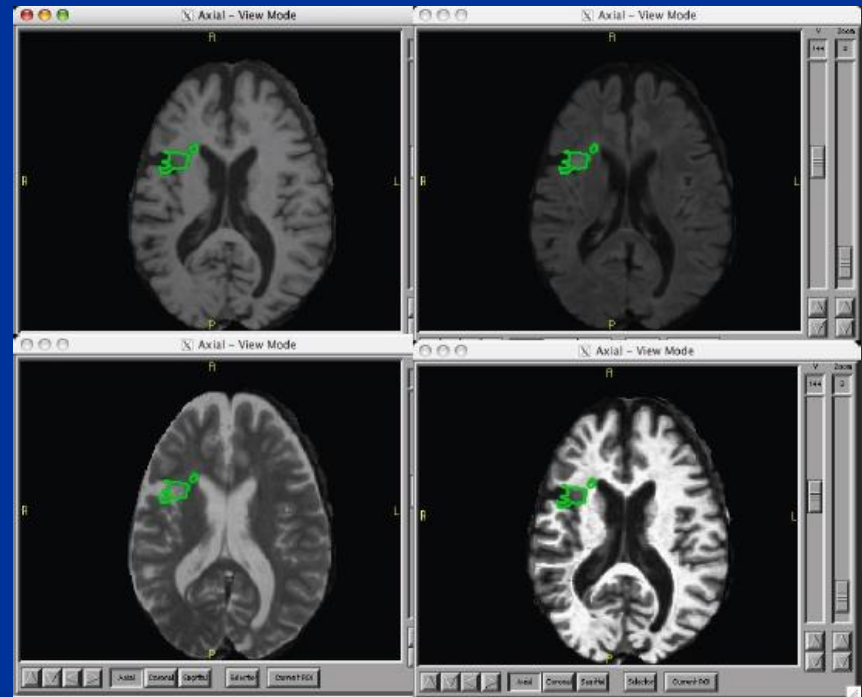
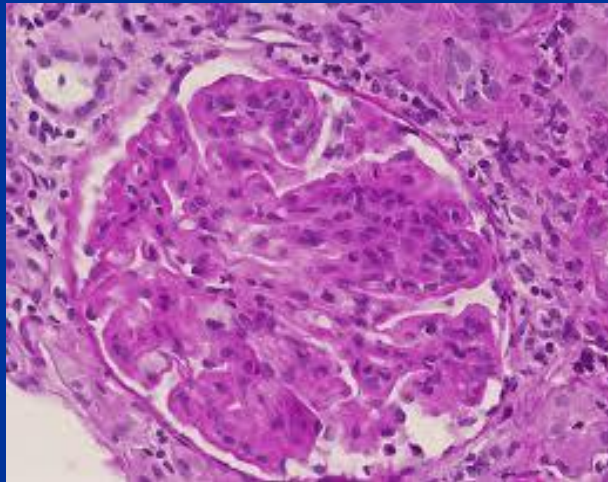


Lupus can involve internal organs



Lupus serious complications

Kidney Failure and Brain damage



Management of Lupus

- Life style: Avoid sunlight, high factor sunscreen, quit smoking
- First step pharmacologic:
 - Nsaids
 - Antimalarials e.g. Hydroxychloroquine for arthritis, rash and malaise
- Corticosteroids- oral or IV depending upon severity
- **Immunosuppressive agents**
 - Oral agents like Azathioprine, Mycophenolate mofetil and Methotrexate
 - IV or oral cyclophosphamide

Scleroderma

- Rare connective tissue disorder, f:m 4:1, onset 30-50 years
- Skin thickening, arthritis and internal organ fibrosis
- Two clinical presentations
 - **Limited cutaneous scleroderma:** hands, feet, lower legs and forearms skin(CREST features)
 - **Diffuse cutaneous scleroderma:** diffuse skin of face, trunk, limbs and internal; organs fibrosis

Clinical features of scleroderma



- Skin thickening
- Heartburn and Reflux
- Difficulty swallowing
- Arthritis
- Raynauds
- Shortness of breath
- Lung scarring
- High pressure on lung arteries

Laboratory findings and investigations

■ Labs:

- high ESR, CRP
- low Hb
- inc creatinine and RBC in Urine
- increased CPK
- low TSH

■ Investigations

- CXR and Esophagogram
- Ct scan chest, ECHO

■ Serology

- +ANA, +anti Scl -70

■ Biopsy: Renal and lung, Skin

Scleroderma Face and hands



Scleroderma complications

Digital vessel narrowing and ulcers, decreased gut motility
and Lung scarring



Management of scleroderma

- Life style: keep body core temperature, avoid smoking
- Pharmacologic treatment: Symptomatic
 - Treat skin infections
 - Calcium channel blockers for Raynuads
 - Proton pump inhibitors e.g. omeprazole for GERD
 - ACE inhibitors for blood pressure
 - Rotating antibiotics for malabsoprtion
- Immunosuppressive agents
 - Prednisone, Hydroxychloroquine and Methotrexate for skin and joints
 - IV cyclophosphamide for lung fibrosis

Sjogren's syndrome

Clinical features

- Inflammatory autoimmune disease affecting exocrine glands
- Ratio f:m 9:1 and onset at age 40-60years
- Clinical features
 - Dryness of exocrine glands
 - Oral Candidiasis and dental caries , gingival recession
 - Chronic esophagitis, Weight loss
 - Sleep disturbance
 - Arthritis, Raynuads and Purpura
 - Nerve damage, Lung scarring

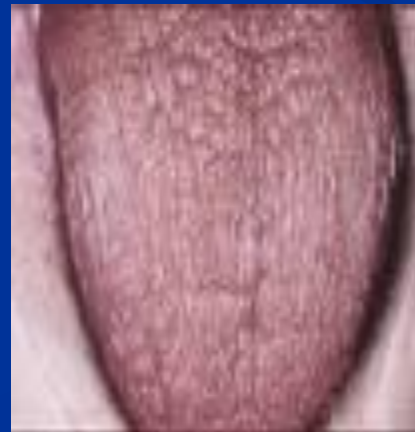
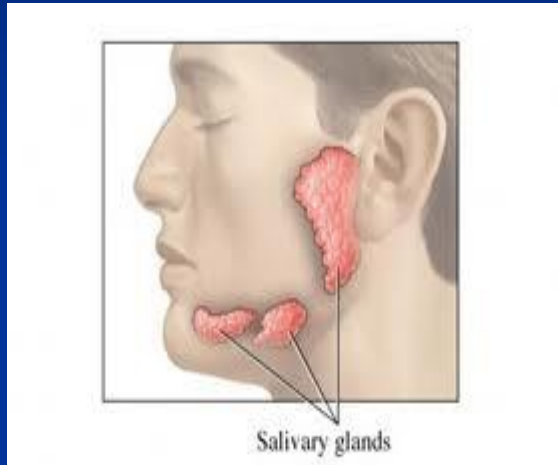
Laboratory findings

Suggestive labs : high ESR, CRP, inc globulins

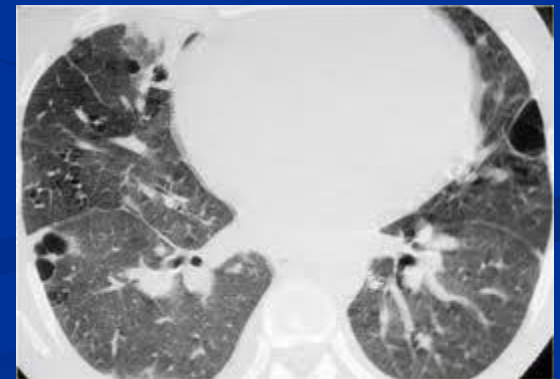
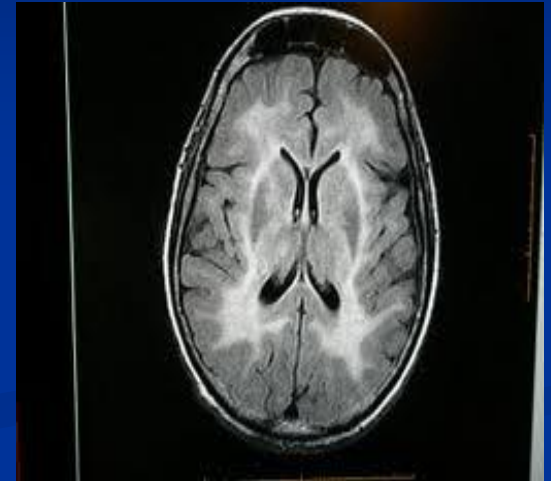
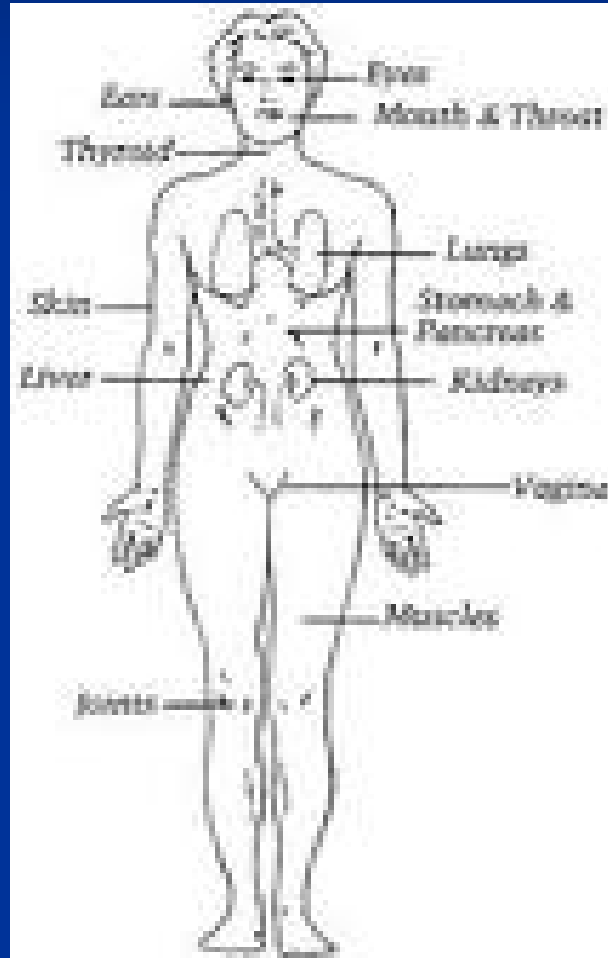
Suggestive Serology: RF, ANA, Anti RO and La antibodies, low c3,c4

Tissue: Minor salivary gland biopsy, lung and kidney biopsy

Signs of Sjogren's syndrome



Complications of Sjogren's syndrome



Management

- Dry eye- artificial tears
- Dry Mouth- sugar free lozenges, good dental care, treatment of oral candidiasis and saliva stimulants e.g. Pilocarpine and Cevimeline
- Arthritis- Nsaids, prednisone
- Immunosuppressive agents: Hydroxychloroquine, Azathioprine and Methotrexate for skin and arthritis
- IV cyclophosphamide for severe lung, brain and kidney disease

Dermatomyositis

- Inflammatory disease of muscle
- Can involve skin in Dermatomyositis
- Clinical features
 - Muscle Weakness in limbs and trunk
 - Rash : heliotrope –purple rash on eyelids, gottrons papules on PIP & MCP, Shawl sign – neck, shoulder girdle
 - Swallowing difficulty
 - Excessive calcium deposits in skin and muscles
 - Lung scarring
 - Can be associated with a undiagnosed cancer

Laboratory findings and Investigations

- Labs
 - inc cpk and aldolase
- EMG
- MRI upper arm or leg
- Serology
- ANA
- Anti-synthetase Jo-1
- Muscle biopsy
- Skin biopsy
- Pan CT scan
- Pelvic ultrasound
- PSA, CA 125

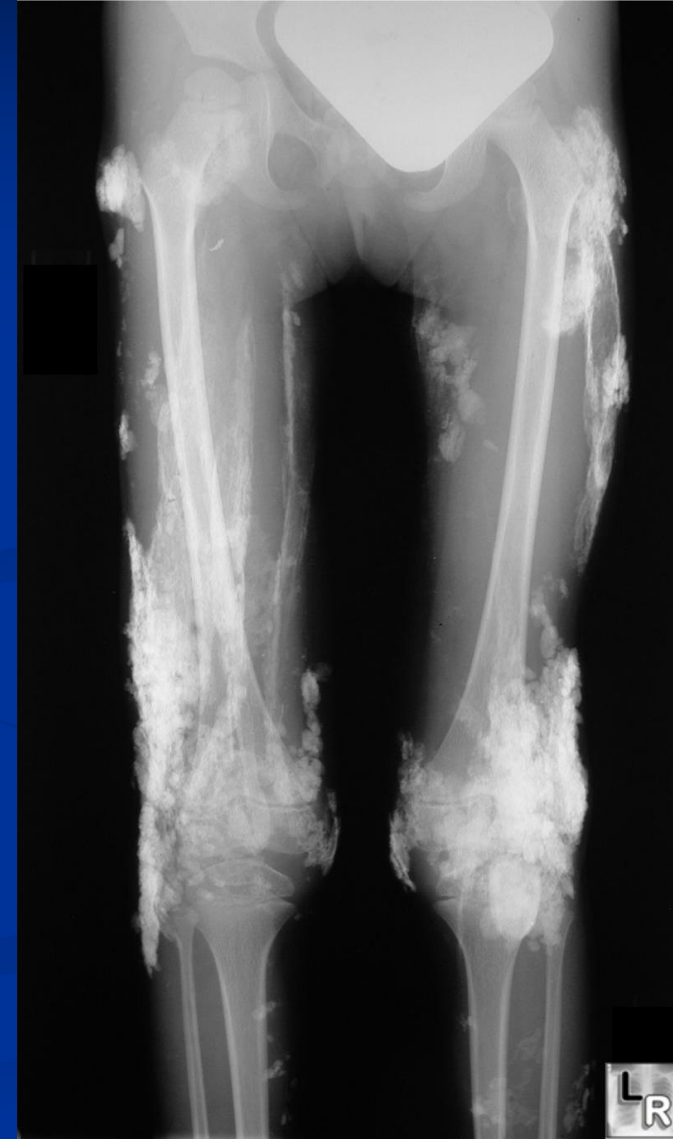
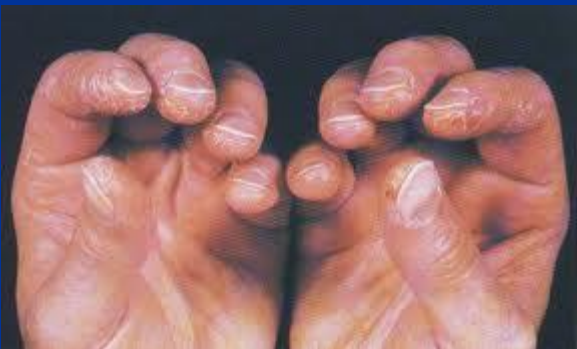
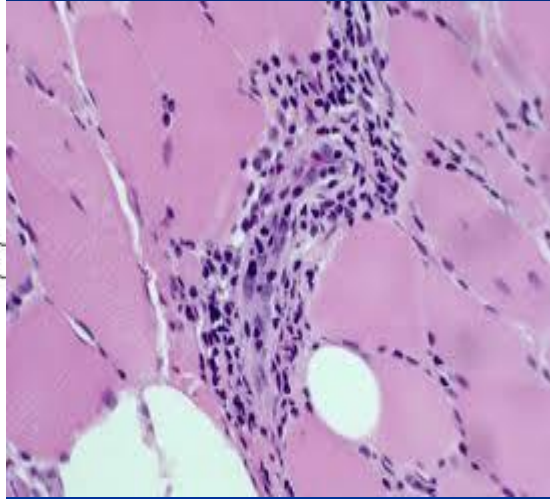
Faces of Dermatomyositis



Signs of Dermatomyositis



Complications of DM



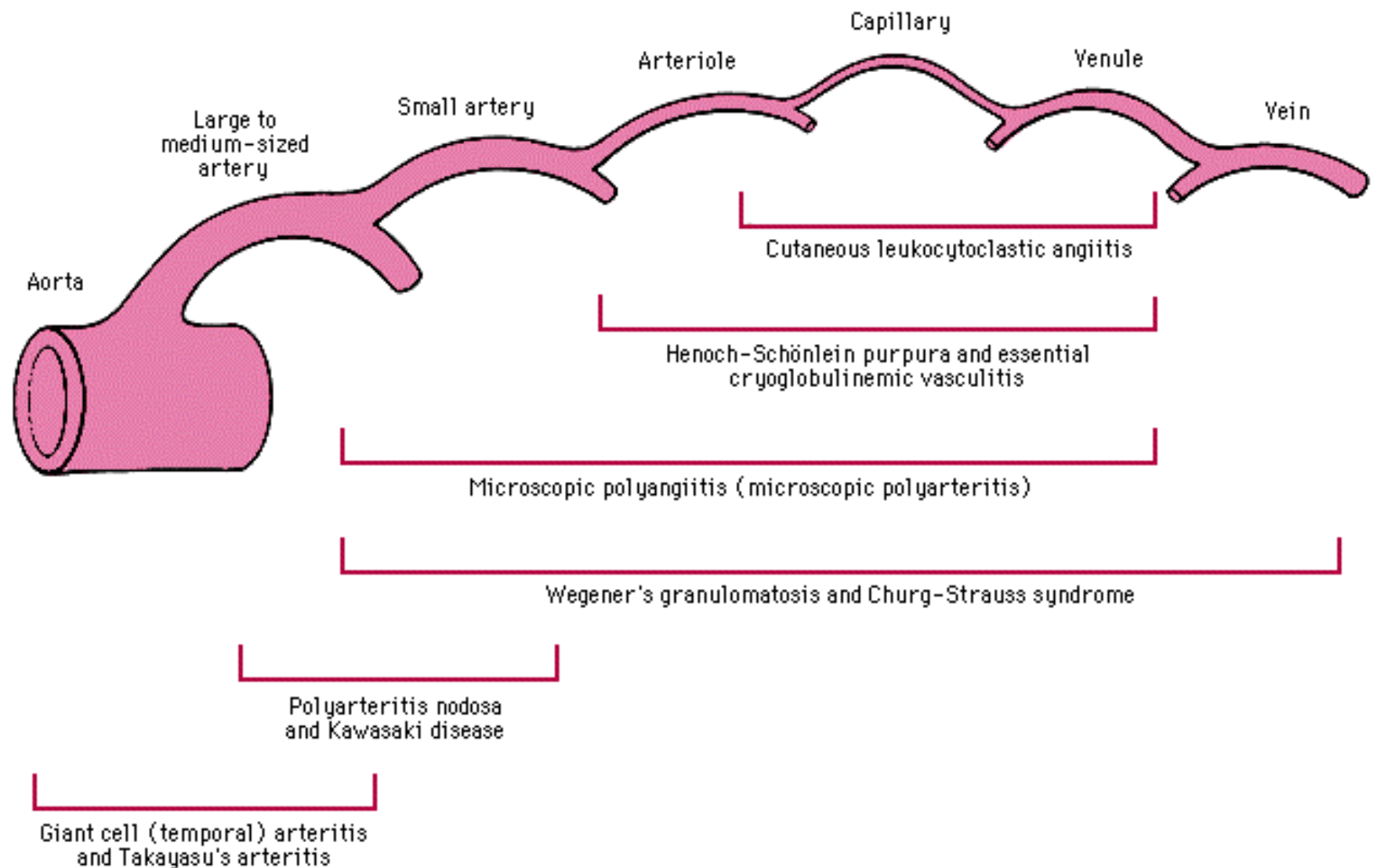
Management

- Need to treat early and aggressive
- Steroids- oral and IV based on severity
- Immunosuppressive agents: Methotrexate, Azathioprine and Mycophenolate mofetil
- IV cyclophosphamide and Rituximab
- IVIG
- Muscle strengthening exercises

Vasculitis

- Uncommon inflammatory disease involving blood vessels
- Most common presentation is cutaneous vasculitis
- Aim of the investigation is to confirm the diagnosis by tissue biopsy
- Exclude infection and malignancy which can mimic vasculitis e.g. atrial myxoma and sub acute endocarditis

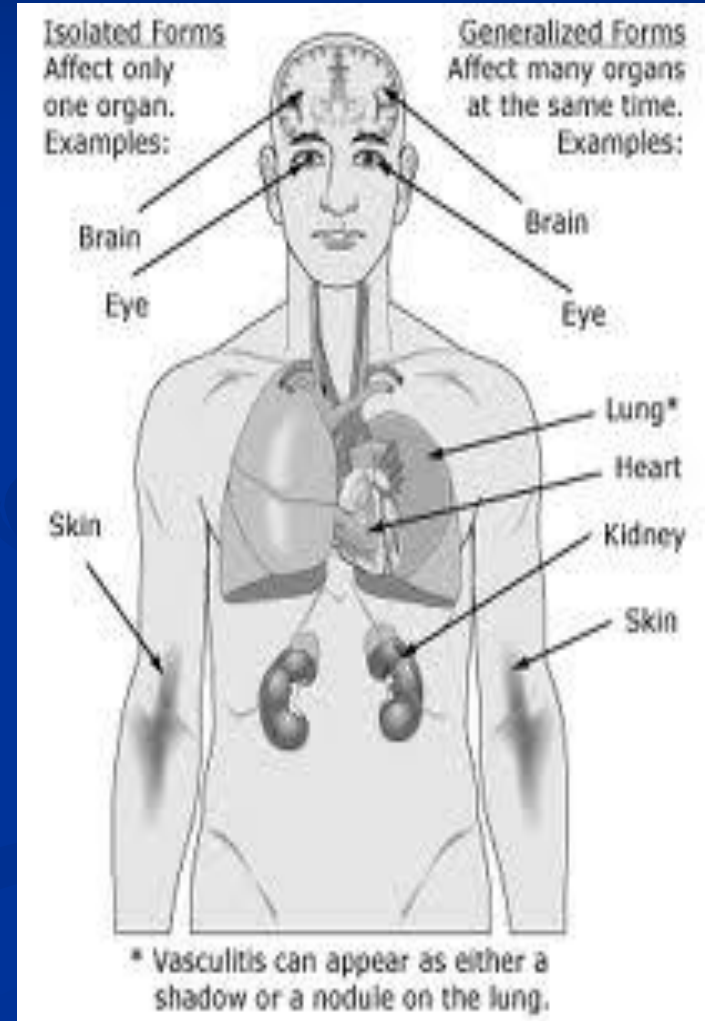
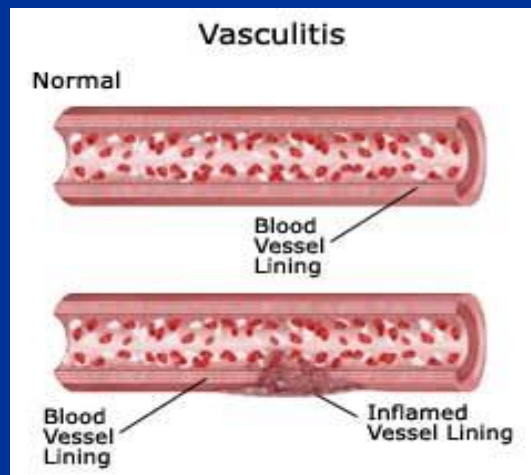
Classification of Vasculitis



Vasculitis

Clinical Features

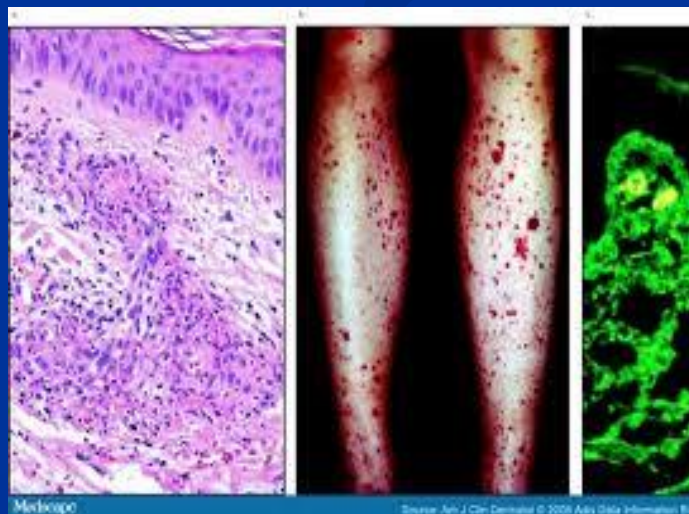
- Skin rash
- Sinusitis
- Kidney failure
- Bleeding from lungs
- Nerve damage
- Vision loss



Laboratory findings and investigations

- Labs: high ESR, CRP
- Low Hb
- RBC and protein in urine
- CPK, Cryoglobulins
- Serology
 - RF, ANA, ANCA
 - MPO and PR3
 - Anti GBM
- Echo
- HRCT chest
- EMG and NC
- Angiogram

Signs of Vasculitis



Complications of Vasculitis

Wegener Granulomatosis



Gingival changes

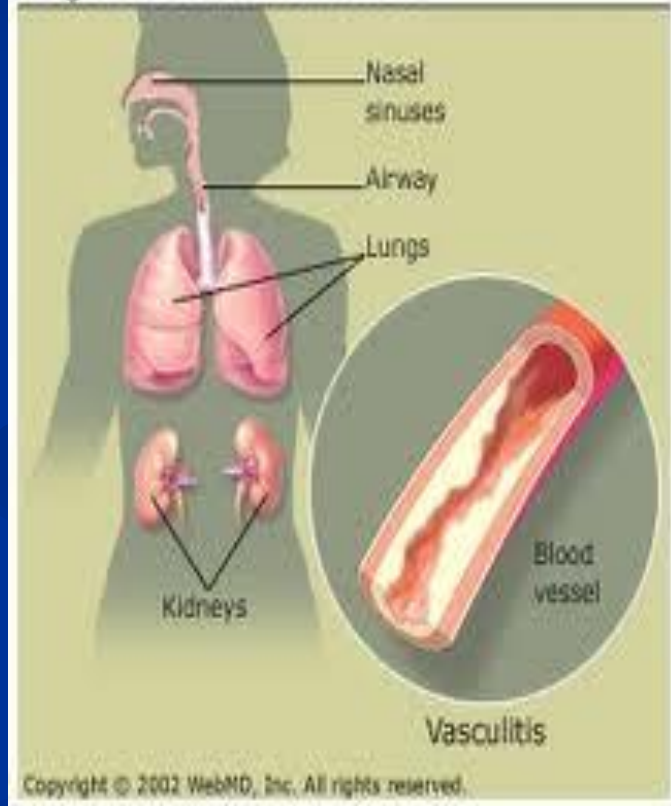


Neutrophil (green) and giant cell (blue) vasculitis

97

© 2007, Michael A. Kahn, DDS, and W. Solomon, DDS

Wegener's Granulomatosis



Management

- Initiate treatment early to induce remission
- IV or oral steroids
- IV and oral cyclophosphamide and Rituximab
- Hydroxychloroquine, Methotrexate, Azathioprine and Mycophenolate mofetil for maintenance of vasculitis or milder forms

Beyond disorder itself

Watch for CTD related complications

- Kidney failure
- Osteoporosis
- High blood pressure
- High cholesterol
- Gastritis
- Infections
- Sterility
- Early menopause
- Cancer
- Depression and Anxiety
- Sexual impairment

Do they have family support

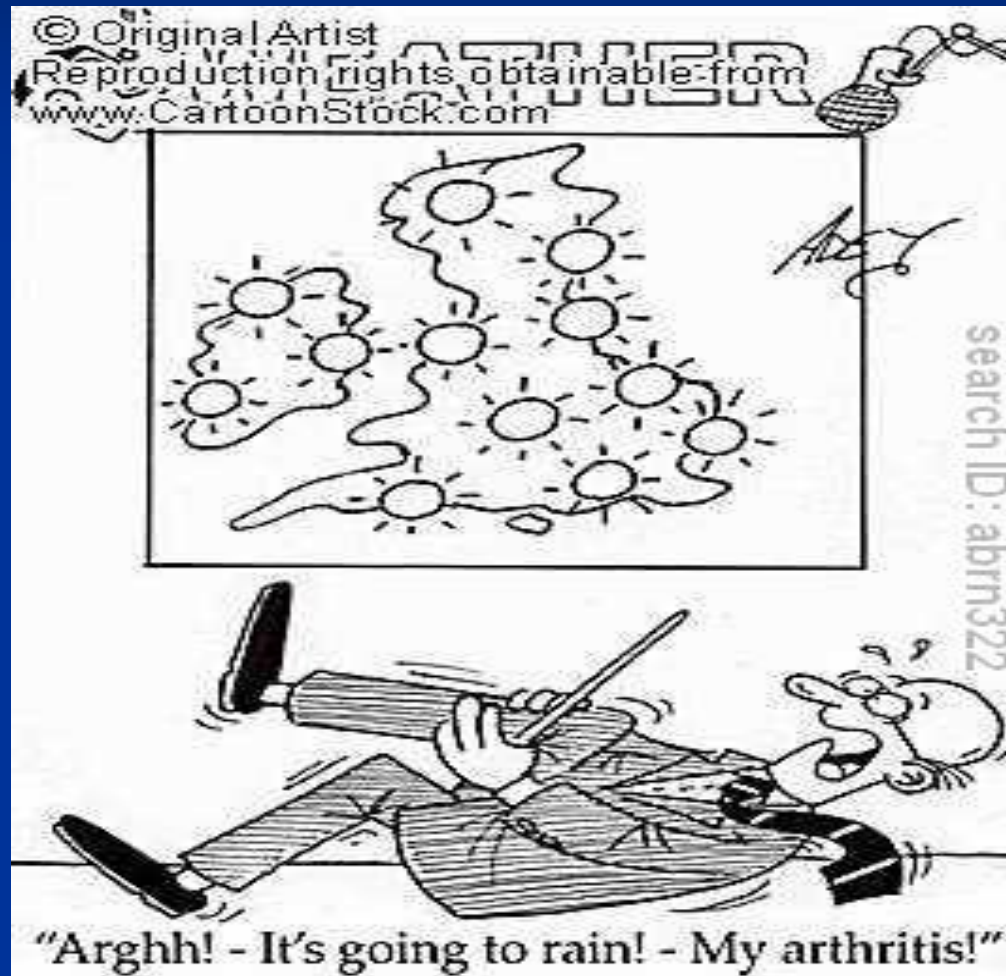


How are their lives affected



"At least your arthritis didn't prevent you from opening that jar of pickles!"

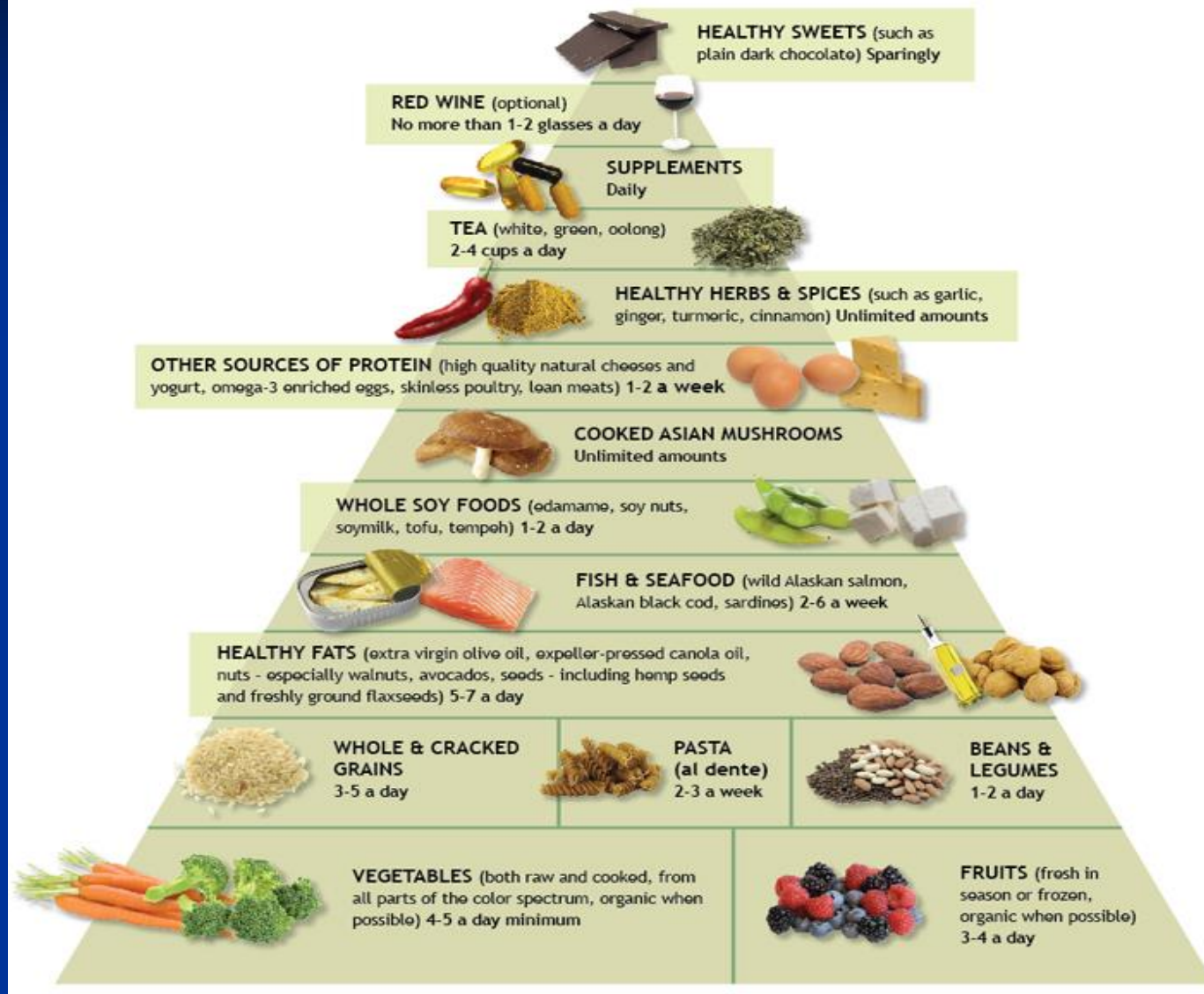
Does weather affect them?



Role of a dietitian

- Adequate calories and nutrients
- Fish oils and plant oils
- There is no diet that can cure rheumatoid arthritis so treatment is necessary
- There are no herbal or nutritional supplements can cure rheumatoid arthritis
- What is an anti-inflammatory diet and what can we suggest
- Dr Weil food pyramid

Dr Weil's food pyramid



Role of regular exercises

- Regular exercise prevents contracture of joints
- Build strong muscle and tendons
- Use of joint protectors and splints
- Rest and stress reduction
- Weight loss is recommended for overweight and obese people

Exercise and Stay Active



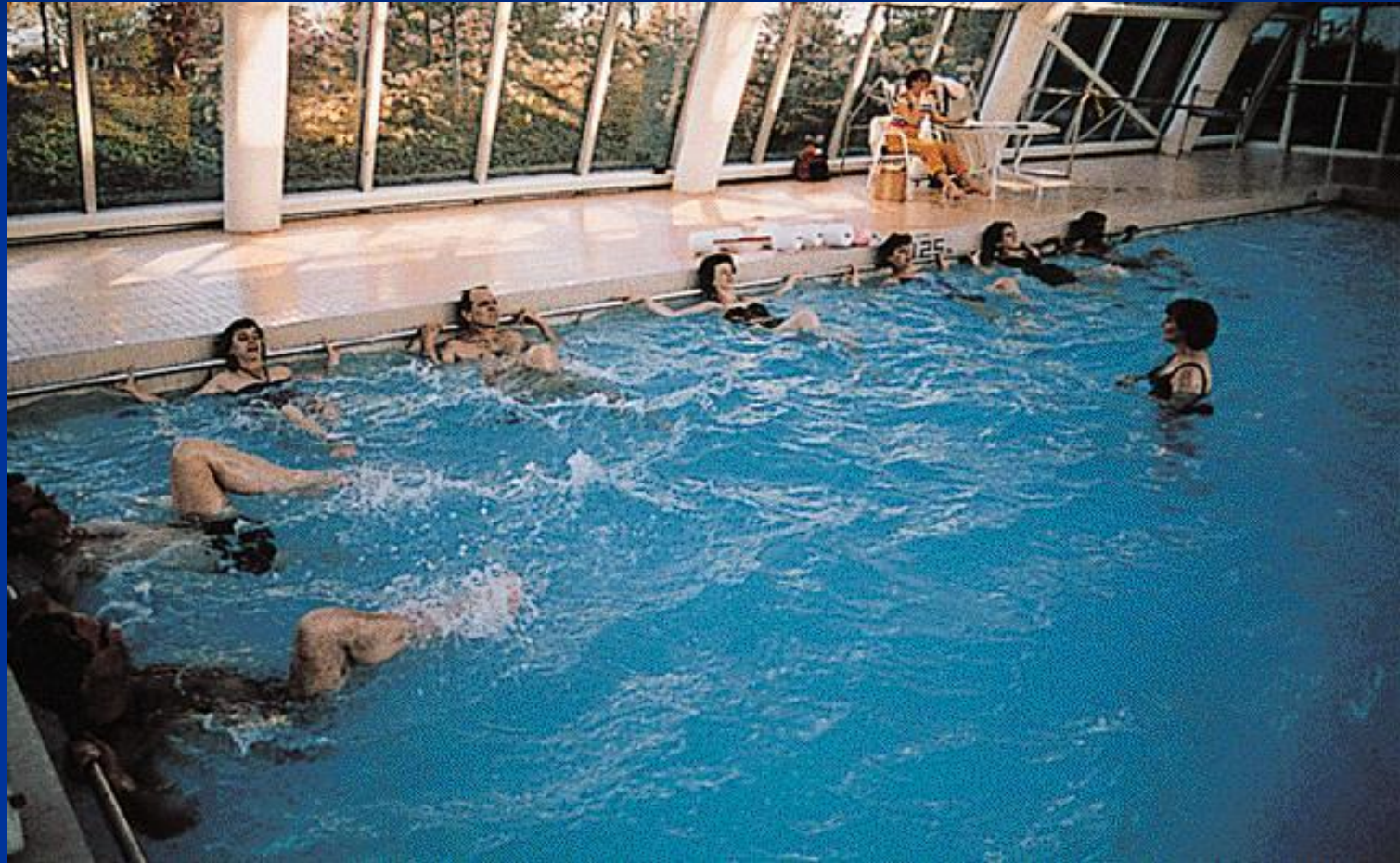
Community Resources



Physical therapy



Swimming and Aquatic therapy





Role of Rheumatologist



- Correct diagnosis and treatment will lead to better results
- Regular visits for assessing progress and avoid complications
- Leading center for cutting edge treatment and research trials
- Personal and trust build up
- Assistance programs for eligible patients

Look for these resources



We all can make a difference

"the prevalence of arthritis could be reduced, at least in part, by greater promotion of effective physical activity, obesity prevention and self-management education programs in local communities."



Our Community, Our Patients, Our Inspiration

Online Resources

- Arthritis Foundation
 - www.arthritis.org
- WebMD
 - <http://arthritis.webmd.com>
- Fibromyalgia
 - www.Knowfibro.com
- Rheumatic Disease Associates
 - www.arthritispa.com
- Institute for Hand and Upper Extremity Rehabilitation
 - <http://www.handinstitute.com>
- Informative web based show for RA
 - www.newwayra.com