# HEY! WANNA COME PLAY IN THE SNOW WITH US?















KEYSTONE

HEAVENLY

NORTHSTAR

KIRKWOOD

# Acute Emergencies and Diagnostic problems in Rheumatology

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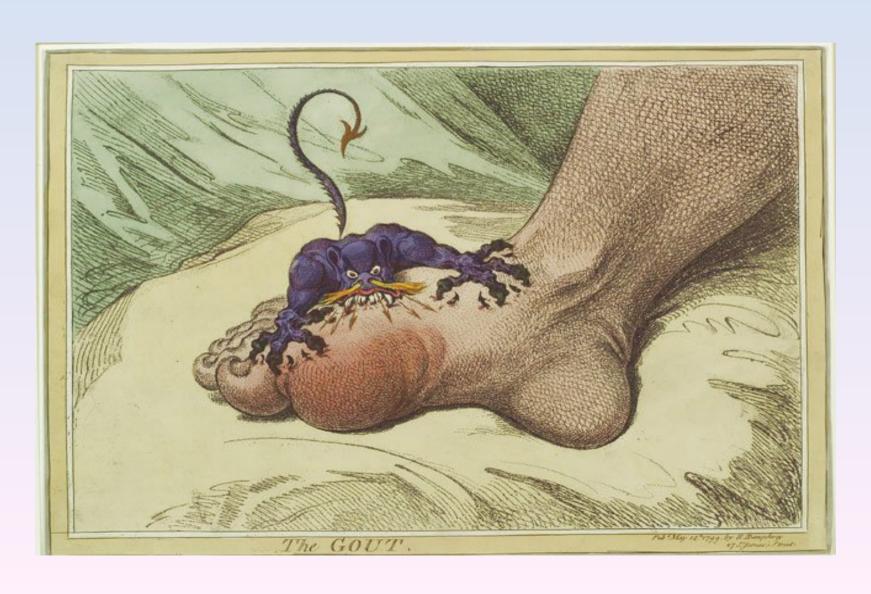
# Acute Rheumatology.

- The Acute Hot joint
- Inflammatory back pain...
- Systemic lupus erythematosus(SLE)
- Inflammatory muscle problems

# Auto antibodies in connective tissue disease

When should we measure autoantibodies?

# The Acute Hot joint!









### Investigations that may help?

History and examination - Pyrexia, Rigors.

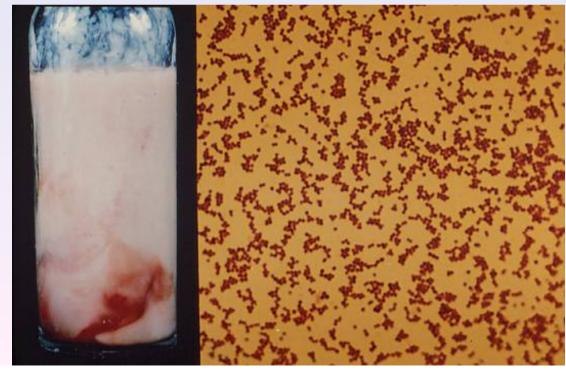
#### **Blood tests**

- Acute phase proteins-CRP/ ESR.
- Raised White cell count
- Serum uric acid
- Blood cultures
- Xrays ./ Ultrasound/ MRI.
- Aspiration of joint

# Should we aspirate this?







# What is the differential Diagnosis?

### **Acute crystal arthritis**

- Urate crystals- gout
- Calcium pyrophosphate
  - Pseudo gout
- Hydroxyapatite
  - Milwaukee shoulder

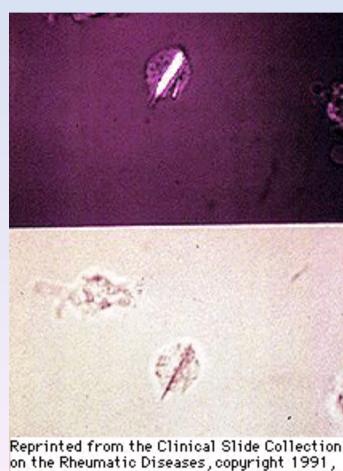
Septic arthritis
Osteomyelitis

New presentation of inflammatory arthritis

# How should it be diagnosed?

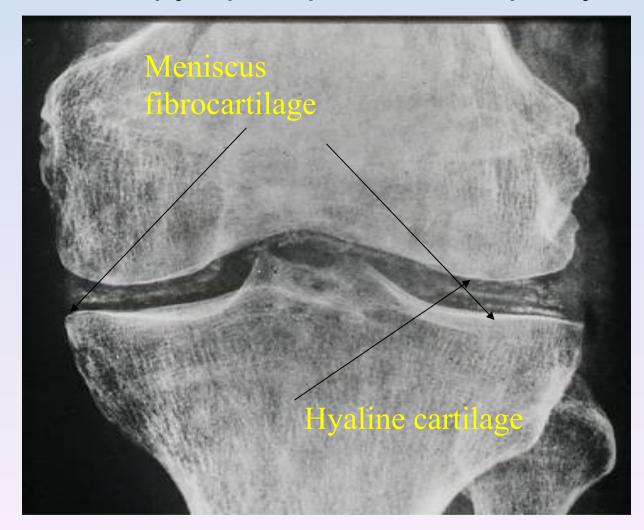
Do we need this?

 Do you know where to send it?



1995, 1997. Used by permission of the American College of Rheumatology.

#### Calcium pyrophosphate arthropathy



CALCIFIED HYALINE AND FIBROCARTILAGE WITH LINEAR AND SPOTTY APPEARANCES AND WELL-PRESERVED JOINT SPACE

# The Xray evidence of Gout

Soft tissue calcification

Subarticular erosion



#### Blood tests often don't help!

- Patient may be pyrexial, and unwell
- Raised CRP, raised ESR
- Raised WCC
- Serum urate may be normal during an acute attack of gout
- Important are Xrays, and aspiration
- Treat as septic arthritis just in case

# **Challenges with the Acute Hot Joint**

- Missing the acute septic arthritis
- Missing the osteomyelitis.
- Crystals not obtained from the joint.

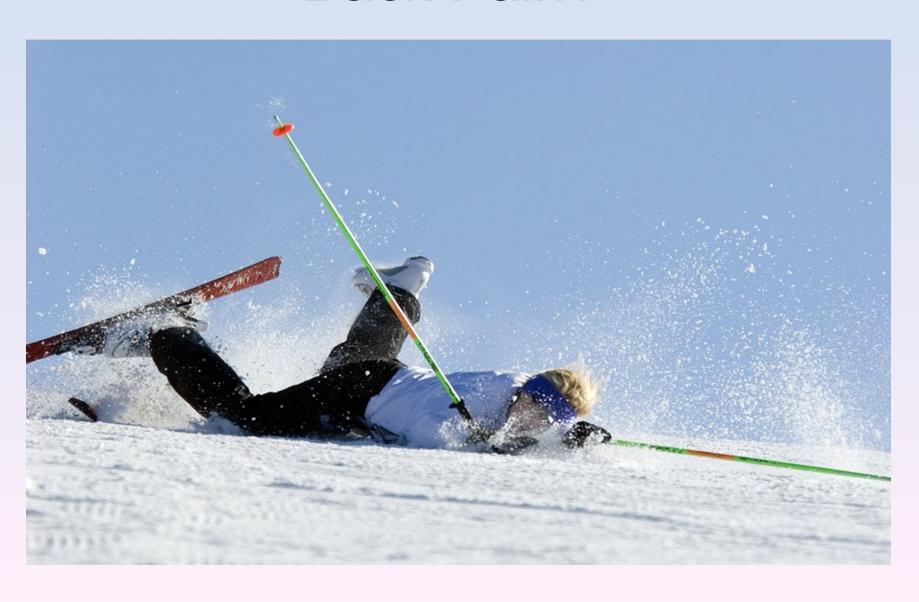
If in doubt treat it as Septic arthritis until you have the aspiration back as clear

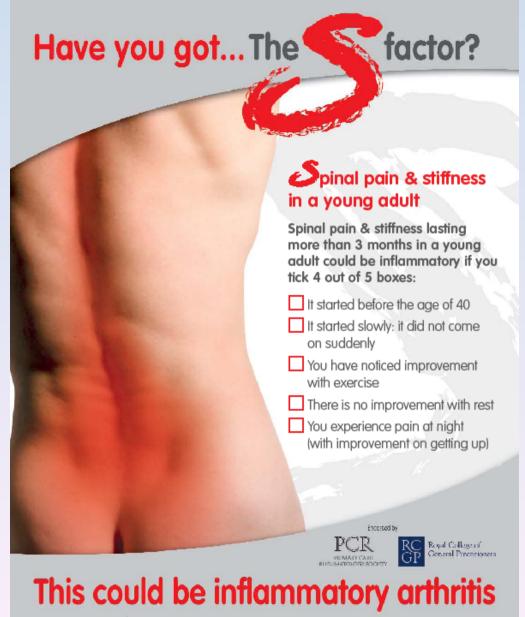
# **Challenges with the Acute Hot Joint**

Diagnosis ??



# Back Pain?



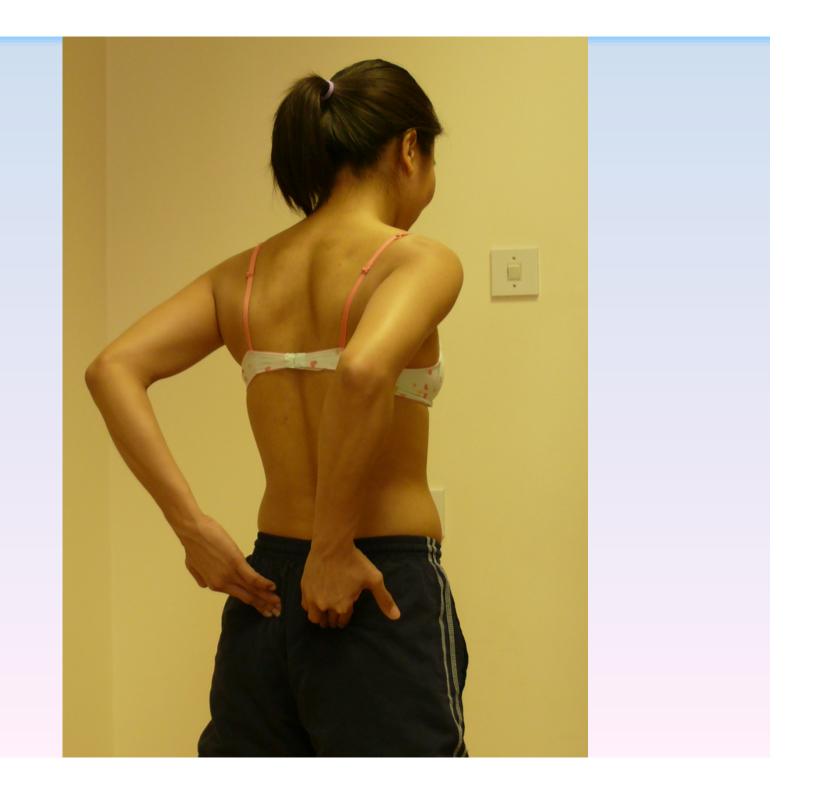


See your doctor now! Delay can cause long term disability

For further information see www.arthritisresearchuk.org



Providing ensures codey and comprow With reorigin tighten and special thoraks to the Phia motiongly Entural Protect Group



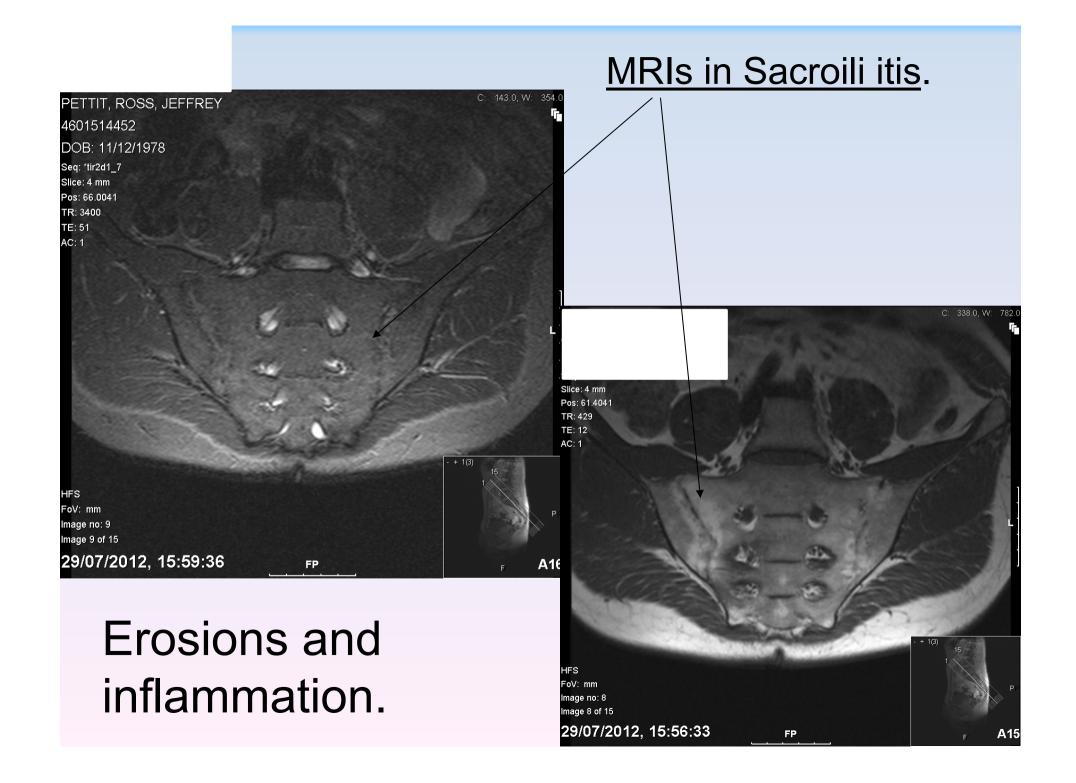
#### Inflammatory back pain

# Back pain of more than 3 months duration is inflammatory if:

- Age at onset less than 40 years
- Insidious onset
- Improvement with exercise
- No improvement with rest
- Pain at night (with improvement on getting up)

The criteria are fulfilled if at least 4 of 5 parameters are present.

ASAS criteria SieperJ et al AnnRheum Dis 2009; 68:784-8

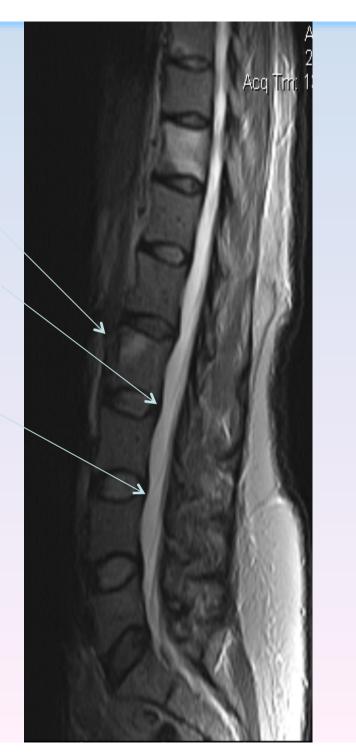


MRI/Xray

Romanus lesions

Syndesmophytes

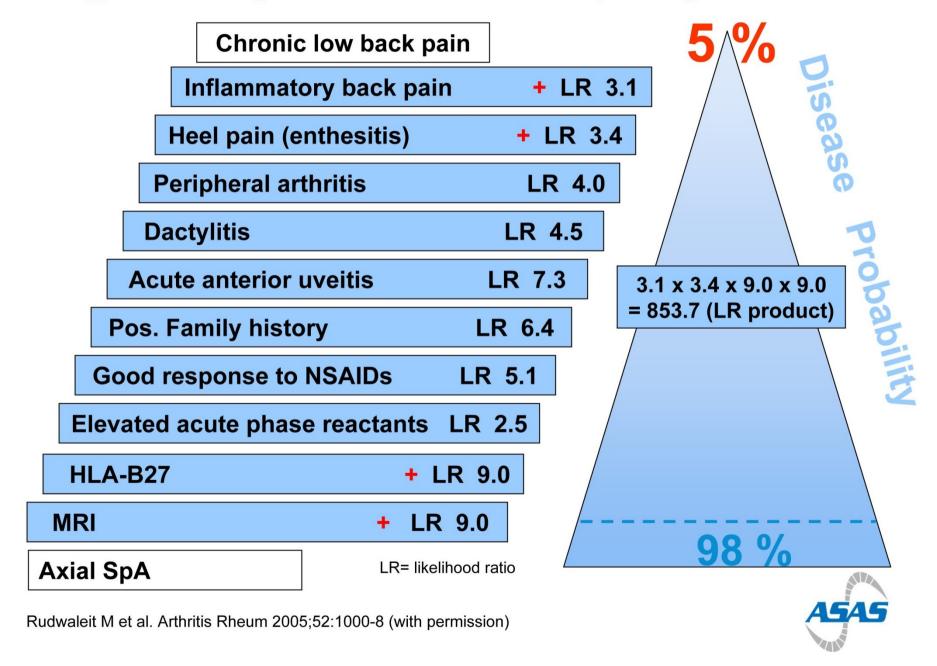
Ankylosis.



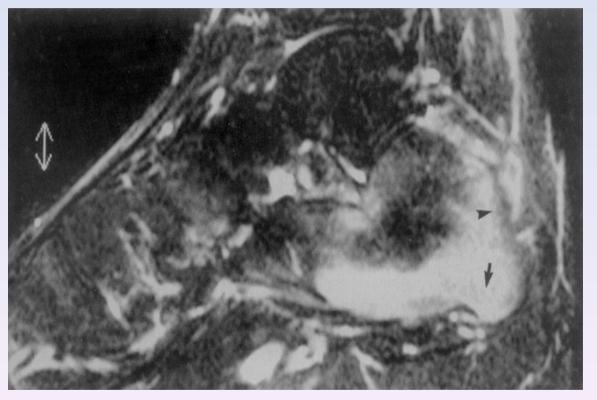


# and 50% of people with AS have other associated problems.....

#### Diagnostic Pyramide for Axial Spondyloarthritis



### Enthesopathy of the heel and tendo achilles.





Marrow oedema and inflammation



#### Inflammatory arthritis of the hip visible on MRI



# Dactylitis – sausage toes,inflammation of the joints and the tendon pulley system

Proximal phalanx



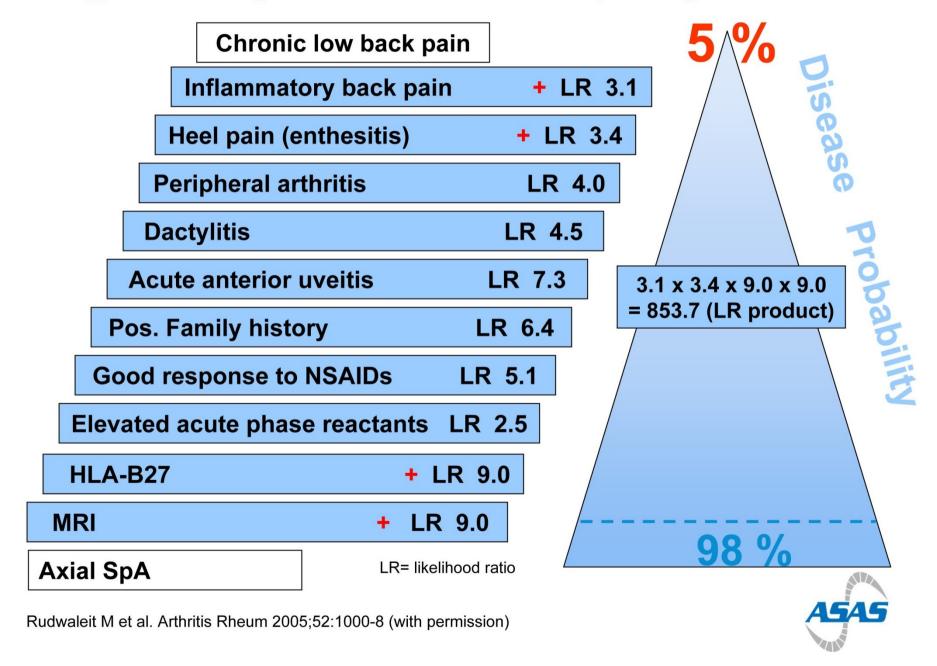
# Acute anterior uveitis, with iritis and Hypopyon.



# Psoriasis in the scalpoften missed.



#### Diagnostic Pyramide for Axial Spondyloarthritis

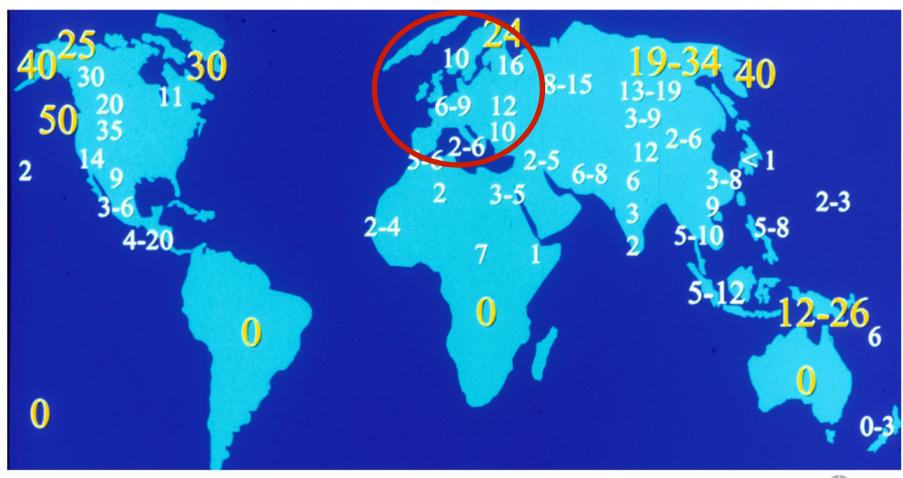


# Spondyloarthritides

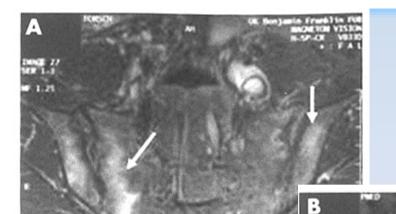
- This group have a similar prevelence to RA
- They share common clinical lesions.
- Inheritance of HLA-B27 is common to all the SpA. The prevalence of these disorders relate to HLA-B27.
- Diagnosis of Ankylosing spondylitis is often delayed
- Identification of inflammatory back pain is very important in determining the diagnosis.
- Use of Anti- tumour necrosis factor biologic drugs has revolutionised the treatment of severe AS.



# Percentage Prevalence of HLA-B27 in Indigenous Populations of the World

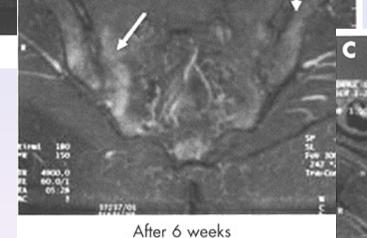






MRI changes improve with Treatment with anti TNF

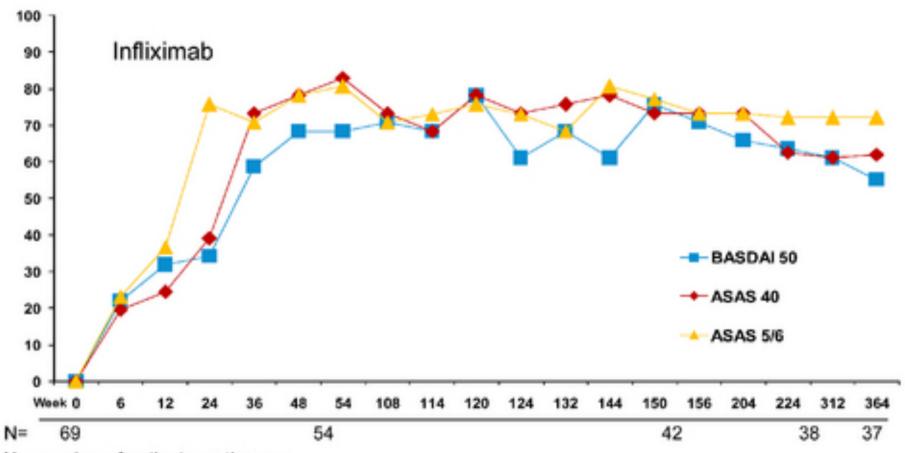




Rudwaleit Met al. Ann Rheum Dis 2005;64:1305



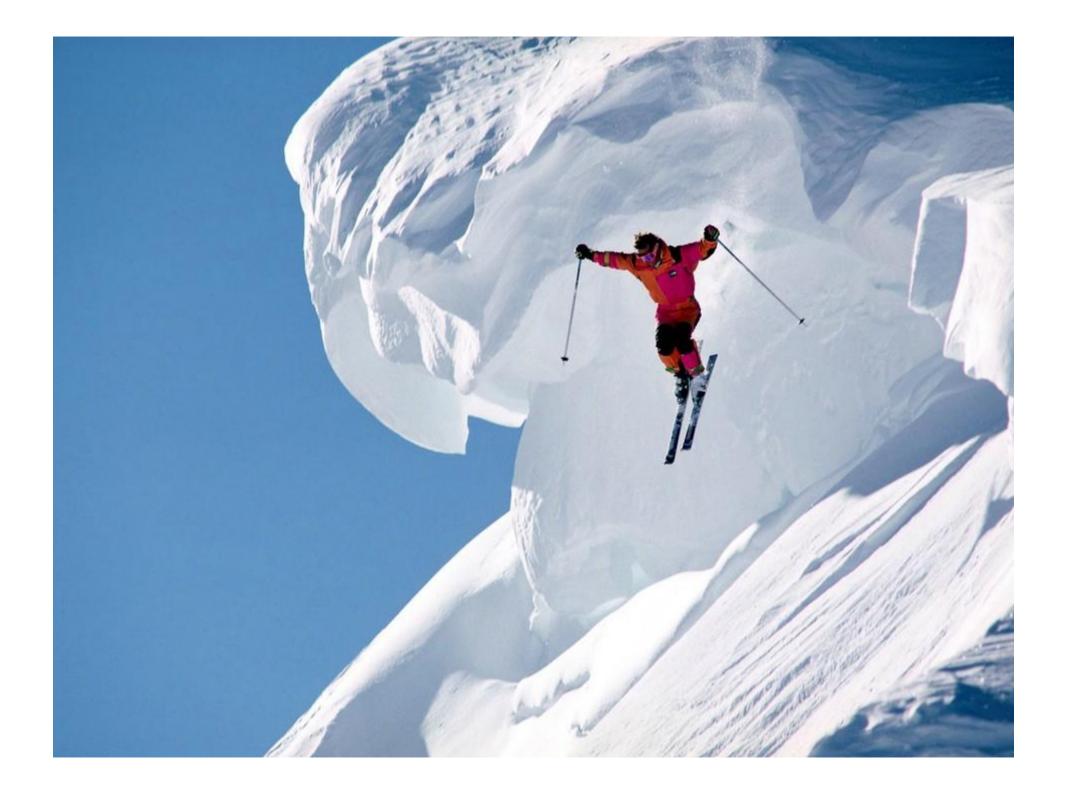
#### Longterm Clinical Efficacy of TNFα-Blocker in AS Results over 7 years



N = number of patients on therapy

Braun J et al. Lancet 2002;359:1187-93 Braun J et al. Ann Rheum Dis 2008;67:340-5 Baraliakos X et al. EULAR 2008, Paris, FRI0290

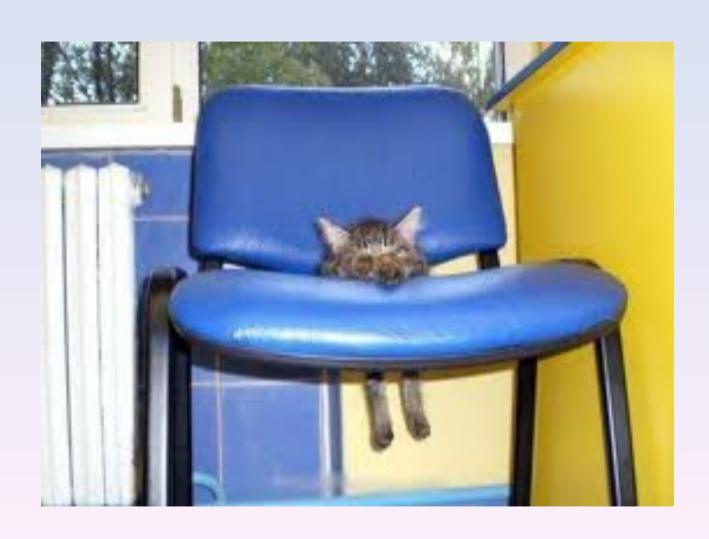




## Connective tissue diseases. Acute presentations



## I can't get out of my chair!

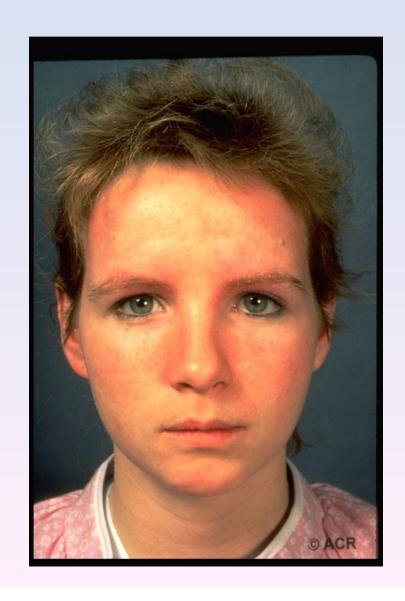


Mrs AU age 35 presented to the dept unable to get out of a chair with severe pain and weakness.

- Facial rash
- Rash on the hands and fingers
- Shortness of breath
- Weakness and muscle pain for 4 weeks
- ?Raynauds syndrome
- Generally unwell.
- Raised ESR, Raised muscle enzymes.
- Raised anti nuclear antibody 1/2500

## Dermatomyositis - facial rashes

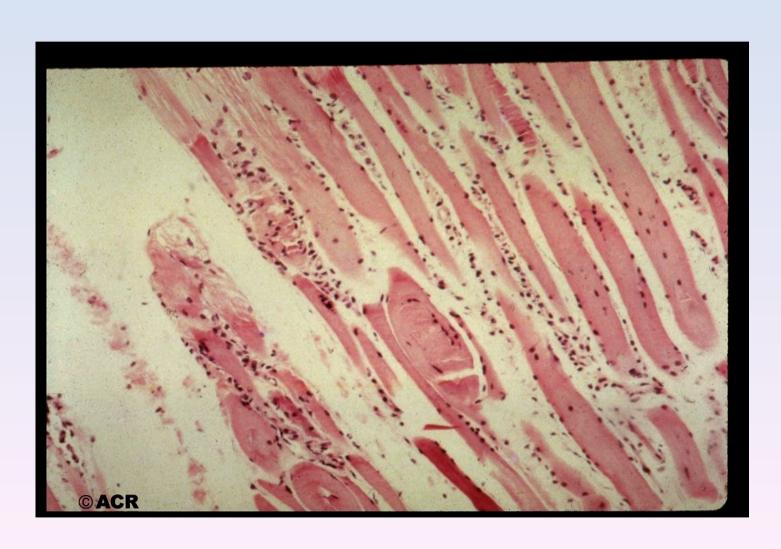




# Dermatomyositis - Gottron's papules



## Acute Myositis



#### **Dermatomyositis**

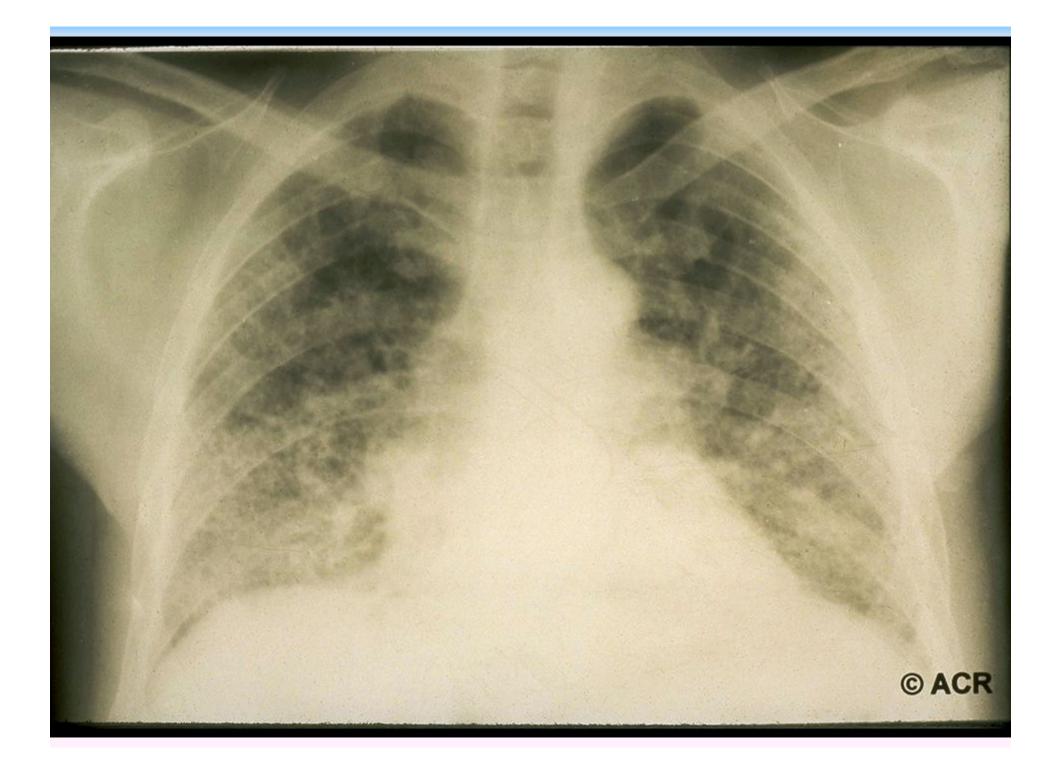
- Rash
- Abnormal muscle enzymes
- Abnormal muscle biopsy and electrical studies and MRI of the thighs.
- Autoantibodies- positive ANA and Extractable nuclear antibodies:--
- Treatment steroids and cyclophosphamide
- ,intravenous Immunoglobulin.
- Rituximab???

#### Myositis and anti synthetase syndrome

- Mechanic hands
- Myositis
- Lung involvement
- Anti Jo1 antibodies
- Other antibodies positive –anti trna synthetase (cytoplasmic antibodies)

### Anti JO 1- mechanic's hands





#### Myositis and anti synthetase syndrome

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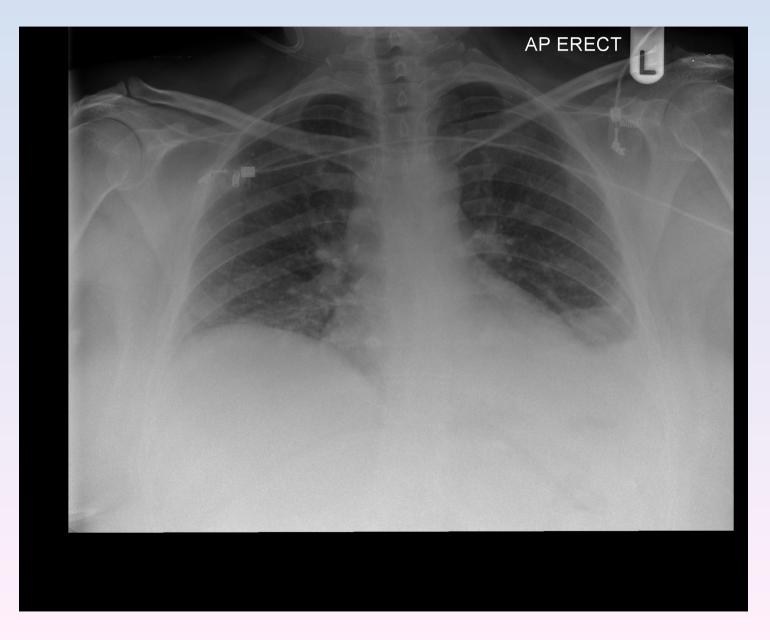
## Connective tissue diseases. Acute presentations



## Case Study AT

- 34yr old vegetarian Asian female
- presented via emergency dept.
- Vomiting, 5kg wt loss, night sweats
- Productive cough & left sided pleuritic chest pain.
- Initial Diagnosis? Pulmonary Tuberculosis? Pneumonia.
- Investigation. Hb 7.8g/dl(low).blood white cells 3.5x10<sub>y</sub>/(low)
- ESR 105mm/hr C reactive protein <4(n).</li>
- O2 saturation 90% on air.

### **Chest Xray**



#### Further history and investigations

Admitted to having been investigated for late miscarriages in her 20s, and being anticardiolipin antibody positive.

Recent general malaise, facial rash.joint pains.

Ct scan showed pulmonary emboli.

Diagnosis –anticardiolipin syndrome and SLE

## CT scan showing embolus



#### AT (2)

- Further investigations.ANA 1/2560 homogenous(very high)
- DNA binding .>300;(HIGH)
- IgG 26.7;IgA 6.9 IgM 2.4;g/l(raised)
- C3 0.3 C4 0.05;g/IComplements(-low)
- Coombs (+) creatinine 105umol/L 24hr urinary protein 2.35 grams/24hrs

- Renal Biopsy WHO Class IV (Diffuse Proliferative Glomurulonephritis)
- Treatment Intravenous Methyl Prednisolone 1gram X3 and anticoagulation
- Dramatic recovery!

## Recognizing SLE

#### "Typical bloods"

- Anaemia
- Lymphopaenia
- Normal/low platelets
- High ESR
- Low C3 and C4
- Autoantibodies



#### Is it a connective tissue disease?

- Raynauds syndrome of recent onset.
- Non specific inflammation with no infection
- Screen for autoantibodies positive.(ANA/DNA)
- Raised ESR with normal C reactive protein.
- Low white cell count with low lymphocyte count and low platelets.
- Rashes.
- Multisystem disease.

#### Who Should I Screen for SLE?

- Fatigue? And generalised muscle pain?
- Is this SLE? Is it fibromyagia?
- The prevelance of SLE is low 40-50 per 100,000.
- Thus the vast majority of people with a low positive ANA do not have SLE( positive predictive value 11%)

## Main symptoms of SLE

- Arthritis 84%
- Malar rash 58%
- Fever 52%
- Photosensitivity 45%
- Nephropathy 39%
- Serositis 36%
- Raynauds phenomenon 34%
- Neurological involvement 27%
- Oral Ulcers 24%

Thrombocytopenia 22%

• ,lymphadenopathy,thrombosis,sicca syndrome < 20%

## Normal Individuals with a positive antinuclear antibodies.

- 1/40 titre 32% normal individuals.
- 1/80 titre -20% normals
- 1/160 titre- 5% normals.
- At the 1/160 dilution :

95% SLE positive

87% Scleroderma

74% Sjogrens syndrome

14% Rheumatoid arthritis

#### **Antinuclear Antibodies**

- Although 95% patients with SLE are ANA pos. the test is only 57% specific. ENAs are more specific.
- Also ANA positive with increasing age especially in women.
- Also relatives of patients with CTD may be positive
- ANA does not reflect disease activity and cannot be used to serially monitor the disease in SLE.

### Tests for SLE if ANA positive

- Anti double stranded DNA antibodies.-97% specific. The titre reflects disease activity/response to treatment.
- Complement C3 and C4-reflect disease activity.
- ESR
- Immunoglobulins
- CRP ? Infection May rise in serositis.

## Available Autoantibody tests to assist diagnosis of connective tissue diseases.

Extractable Nuclear antibodies (ENAs)

**Sm** in 15% patients with **SLE**, high specificity –neurolupus.

**SS-A/ Ro** 70 % of Sjogrens syndrome—low specificity . 35% of patients with **SLE**.

Consorted boost block

Congenital heart block.

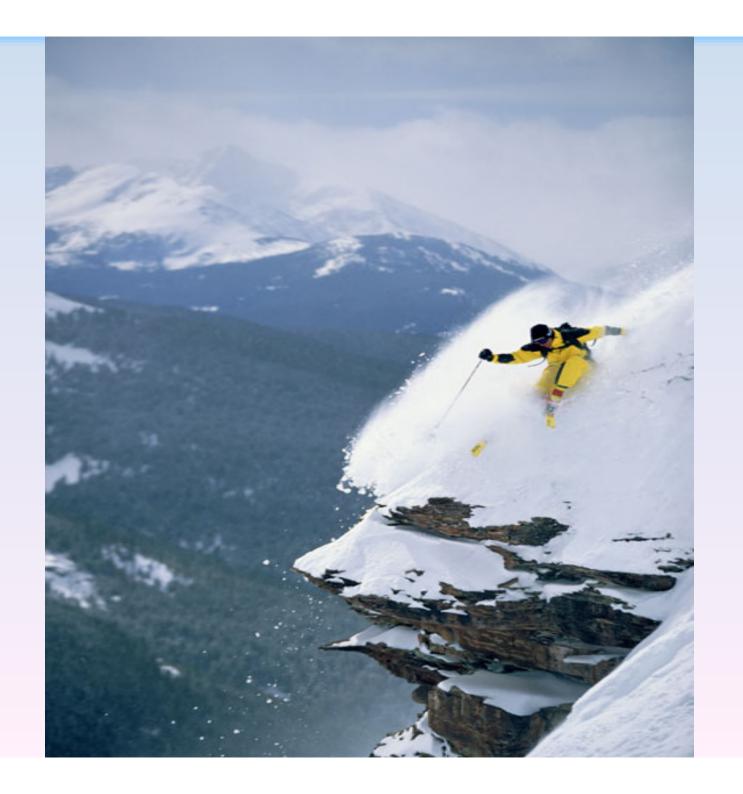
SS-B/La- 40% Sjogrens and 15% SLE.

ScI-70 in 15% scleroderma high specificity- lung fibrosis. antiCentromere in 25% scleroderma mod-specificity-skin

**Jo**-1, anti synthetase abs.very specific + lung fibrosis and Polymyositis

## Northwick park hospital Harrow





## Raynauds



### Small vessel ischaemia



