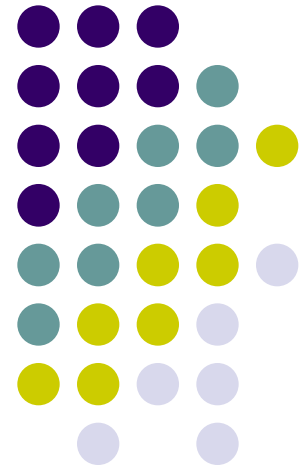
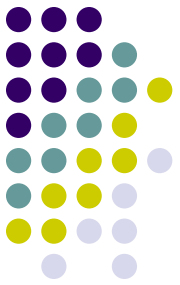


Giant Cell Arteritis

Dr Thirau Marianayagam

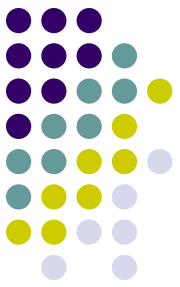
Consultant Rheumatology





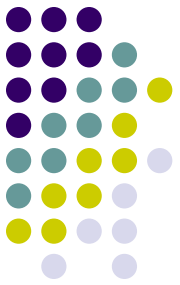
Polymyalgia Rheumatica

- Inflammatory condition of the muscles & joints
- Causes pain and stiffness
- Limb girdle fashion
- Constitutional symptoms
- Fatigue, fever
- 20% can develop concomittant GCA



Signs & symptoms

- Age >60
- shoulder pain on both sides
- morning stiffness that lasts at least 45 minutes
- Elevated CRP (or ESR if you must)
- new bilateral hip pain
- no swelling in the small joints of the hands and feet
- no evidence of rheumatoid arthritis, such as swollen joints or positive blood tests



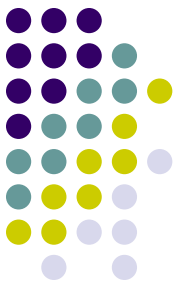
Differential Diagnosis

- Active malignancy
- Infection
- Other inflammatory rheumatic diseases
- Drug-induced myalgia
- Chronic pain syndromes
- Endocrine disease
- Neurological conditions, e.g. Parkinsons disease

Baseline investigations



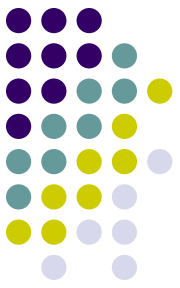
- Full blood count
- CRP +/- ESR
- Urea and electrolytes
- Liver function tests
- Bone profile
- Protein electrophoresis (plus urinary Bence Jones Protein)
- Thyroid stimulating hormone
- Creatine kinase
- RF (ANA and anti-CCP antibodies may be considered)
- Dipstick urinalysis
- Chest X-ray may be required



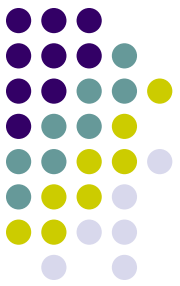
Treatment – steroids

- **Prednisolone 15mg OD**
- A patient-reported global improvement of 70% within a week of commencing steroids is consistent with PMR, with normalization of inflammatory markers in 4 weeks. A lesser response should prompt the search for an alternative condition.
- The diagnosis of PMR should be confirmed on further follow-up. Follow-up visits should include vigilance for mimicking conditions.

Steroid weaning programme



- prednisolone 15 mg OD for 3 weeks
- Then 12.5 mg for 3 weeks
- Then 10 mg for 4–6 weeks
- Then reduction by 1 mg every 4–8 weeks or alternate day reductions (e.g. 10/7.5 mg alternate days, etc.)
- Aim for 0mg
- Bone protection/ PPI/ DM & BP reviews
- Treatment should be complete within 1-2 years



Treatment programme

- Follow-up schedule:
Weeks 0, 1–3, 6, Months 3, 6, 9, 12 in first year (with extra visits for relapses or adverse events).
- Bloods (FBC, U&E, glucose, CRP)

At each visit, patients should be assessed for the following:

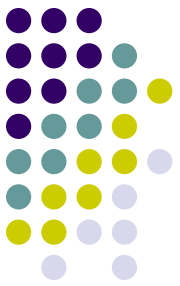


- Response to treatment: proximal pain, fatigue and morning stiffness It is important to distinguish between symptoms due to inflammation and those due to co-existing degenerative problems.
- Complications of disease including symptoms of GCA, e.g. headaches, jaw claudication and large-vessel disease
- Steroid-related adverse events
- Atypical features or those suggesting an alternative diagnosis

When to refer to rheumatology



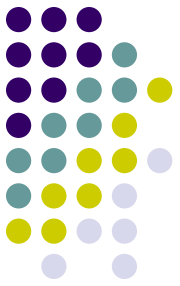
- Atypical features or features that increase likelihood of a non-PMR diagnosis:
- Age <60 years
- Chronic onset (>2 months)
- Lack of shoulder involvement
- Lack of inflammatory stiffness
- Prominent systemic features, weight loss, night pain, neurological signs
- Features of other rheumatic disease
- Normal or extremely high acute-phase response



Treatment dilemmas

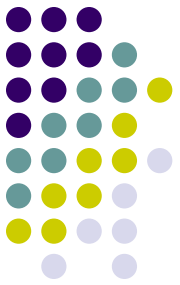
- Incomplete, poorly sustained or non-response to corticosteroids
- Inability to reduce corticosteroids
- Contraindications to corticosteroid therapy
- The need for prolonged corticosteroid therapy (>2 years)

Consider DMARD therapy after two relapses



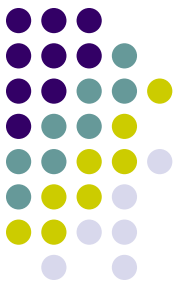
Summary

- What is GCA?
- History
- Signs & Symptoms
- Investigations
- Treatment
- Complications



What is GCA?

- Large & medium vessel vasculitis often affecting the cranial arteries
- Annual incidence of 15-25 per 100 000
- Almost exclusively seen above age 60 yrs
- More prevalent in women



History

- Usually gradual but can be abrupt onset of symptoms
- Headache 68%
 - Often but not exclusively unilateral
- Jaw Claudication 50%
- Transient visual symptoms 16%
- Fixed visual symptoms 14%
- CNS abnormalities

History Cont

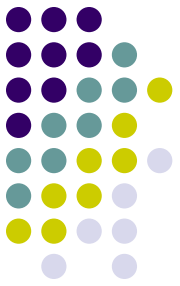


- Swallowing claudication/ dysphagia 8%
- Tongue Claudication 6%
- Limb Claudication 4%

Constitutional Symptoms



- Fatigue
- Widespread myalgia/ PMR (50%)
- Fevers
- Night sweats
- Anorexia
- THEREFORE NEED TO CONSIDER MALIGNANCY



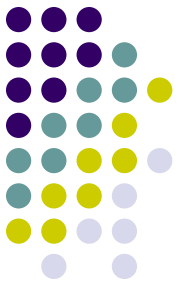
Clinical Signs

- Weight loss & anorexia 50%
- Reduced temporal artery pulsations 46%
- Fever 42%
- Arterial tenderness 27%
- Erythematous / swollen scalp arteries 23%
- Fundoscopic abnormalities 18%

American College of Rheumatology

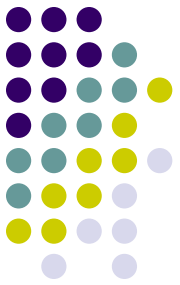


- Age at disease onset >50 years
- New headache: new onset or type of localised pain in the head
- Temporal artery abnormality
- ESR>50/ CRP>40 NB
 - CRP is more sensitive & easier to interpret
- Abnormal artery biopsy
 - Presence of 3 or more yields a sensitivity of 93.5%



Investigations - bloods

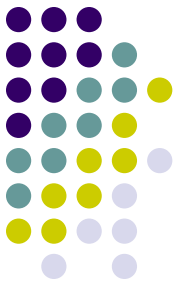
- ESR >50
- CRP
- Mild anaemia
- Abnormal LFTs
- Thrombocytosis
- ANCA



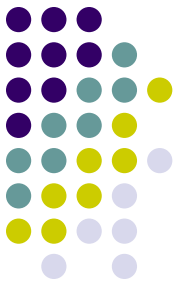
Investigations cont.

- CXR
 - Rule out underlying malignancy
 - Aortic aneurysm
- Temporal artery biopsy
 - Needs to be done ASAP
 - Book via the vascular surgeons
 - 85% sensitive
 - 100% specific
- Urine analysis

Treatment



- Steroids
 - 1mg/kg/day for 2-4 weeks
 - If symptoms stable reduce by 20mg every 2 weeks to 20mg OD
 - Reduce by 2.5mg every 2 weeks to 10mg OD
 - Reduce by 1 mg every month to zero
 - Steroid dose governed by signs & symptoms
- Steroid counselling including steroid card



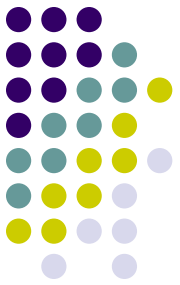
Treatment

- Bone protection
 - Calcium/ vitamin D supplements
- Anticoagulation
 - Aspirin 75mg OD
- PPI/ gastric protection



Complications

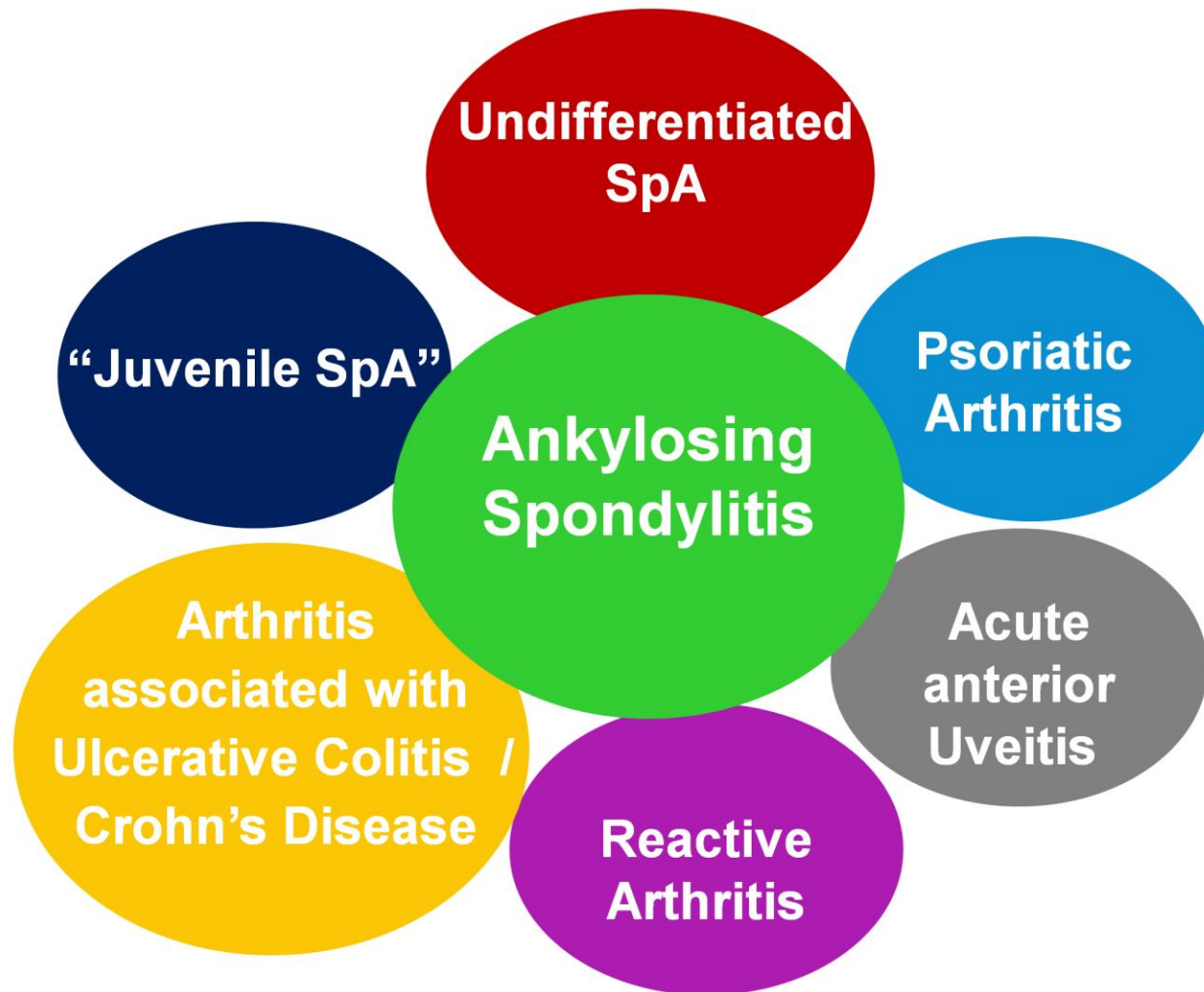
- Visual Loss
- Tongue necrosis
- Limb claudication
- Drug Side effects
 - Steroids
 - Bisphosphonates
 - PPI



Any Questions

- Further information
- Arthritis Research UK
- <http://www.arthritisresearchuk.org/arthritis-information/conditions/giant-cell-arteritis.aspx>
- British Society of Rheumatology GCA Guidelines
- http://www.rheumatology.org.uk/includes/documents/cm_docs/2010/m/2_management_of_giant_cell_arteritis.pdf

Spondyloarthritides (SpA)



The Incidence of Ankylosing Spondylitis

- The annual incidence of AS requiring antirheumatic medication was 6.9 per 100,000 adults (95% CI = 6.0 to 7.8) in Finland¹.
- The incidence of AS was shown to be 7.26 per 100,000 inhabitants in northern Norway².
- The overall age- and sex-adjusted incidence of AS was 7.3 per 100,000 person years (95% CI = 6.1 to 8.4) in the U.S³.

No data available for axial spondyloarthritis (proportion AS/nr-axSpA about 50/50).

1. Kaipiainen-Seppänen O et al. J Rheumatol 1997;24:496-499
2. Bakland G et al. Arthritis Rheum 2005;53:850-855
3. Carbone LD et al. Arthritis Rheum 1992;35:1476-82



Acute Arthritis of the Right Knee in a Patient with Peripheral Spondyloarthritis



Enthesitis (Insertion of Achilles Tendon at Calcaneus) Right Heel

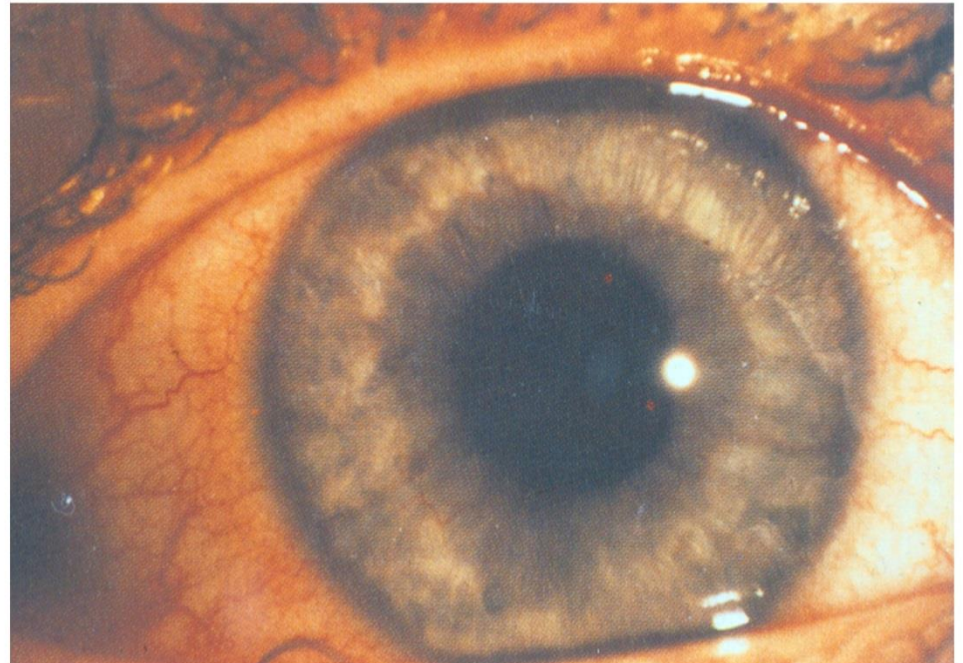


Skin Manifestations in Spondyloarthritis

- Psoriasis
- Erythema nodosum
- Pyoderma gangrenosum
- Keratoderma blenorrhagicum

Eye: Acute Anterior Uveitis in Spondyloarthritis

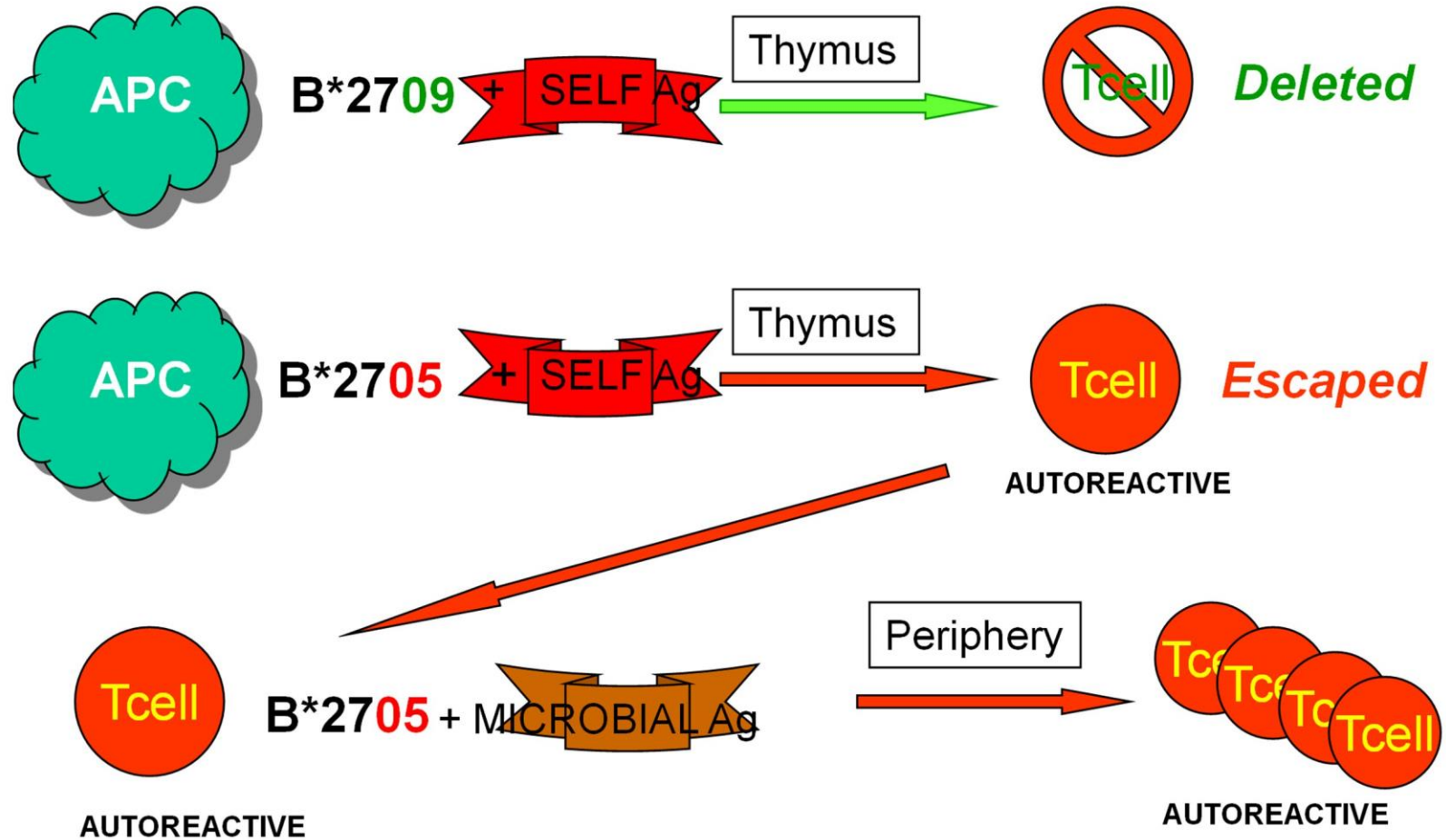
- Acute onset
- Unilateral
- Anterior
- Spontaneous remission
- Recurrent
- Related to HLA B27



AS Patient with no Flexibility of the Lumbar Spine on Bending Forward (Flat Back)



Possible Role of HLA-B27 in the Pathogenesis of Spondyloarthritis



ASAS Classification Criteria for Axial Spondyloarthritis (SpA)

In patients with ≥ 3 months back pain and age at onset < 45 years

Sacroiliitis on imaging*
plus
 ≥ 1 SpA feature

OR

HLA-B27
plus
 ≥ 2 other SpA features

***Sacroiliitis on imaging**

- active (acute) inflammation on MRI highly suggestive of sacroiliitis associated with SpA
- definite radiographic sacroiliitis according to the modified New York criteria

SpA features:

- inflammatory back pain
- arthritis
- enthesitis (heel)
- uveitis
- dactylitis
- psoriasis
- Crohn's/colitis
- good response to NSAIDs
- family history for SpA
- HLA-B27
- elevated CRP

n=649 patients with back pain;

Overall

Sensitivity: 82.9%, Specificity: 84.4%

Imaging arm alone

Sensitivity: 66.2%, Specificity: 97.3%

Clinical arm alone

Sensitivity: 56.6%, Specificity: 83.3%



Inflammatory Back Pain Criteria (Calin)

Ankylosing Spondylitis n=42; mechanical low back pain n=21

- age at onset < 40 years
- duration of back pain > 3 months
- insidious onset
- morning stiffness
- improvement with exercise

Sensitivity: 95%; Specificity: 76%

Inflammatory back pain if 4/5 criteria are present.

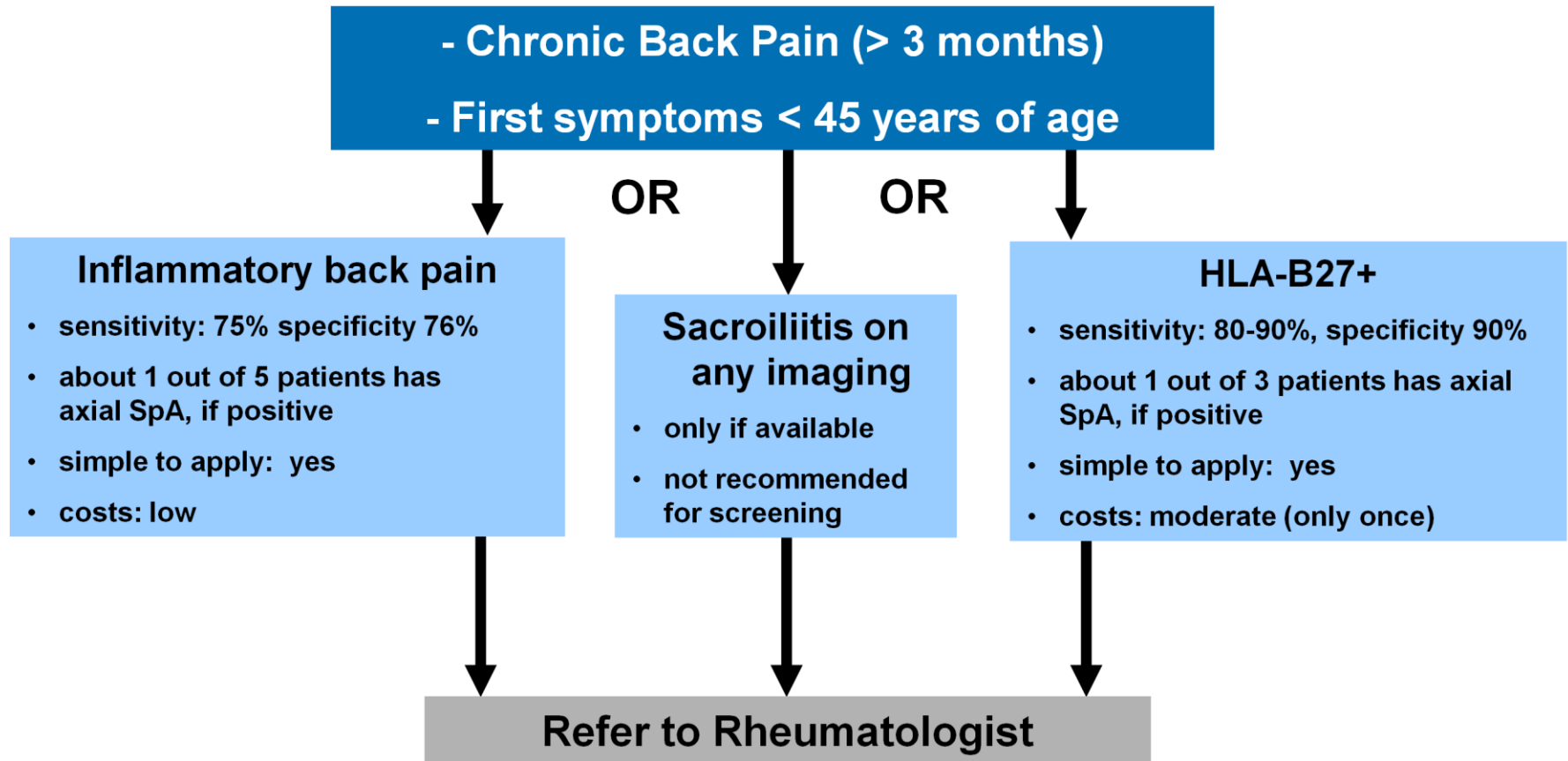
ASAS Inflammatory Back Pain Criteria by Experts (Chronic Back Pain; n=648)

- age at onset < 40 years
- insidious onset
- improvement with exercise
- no improvement with rest
- pain at night (with improvement upon getting up)

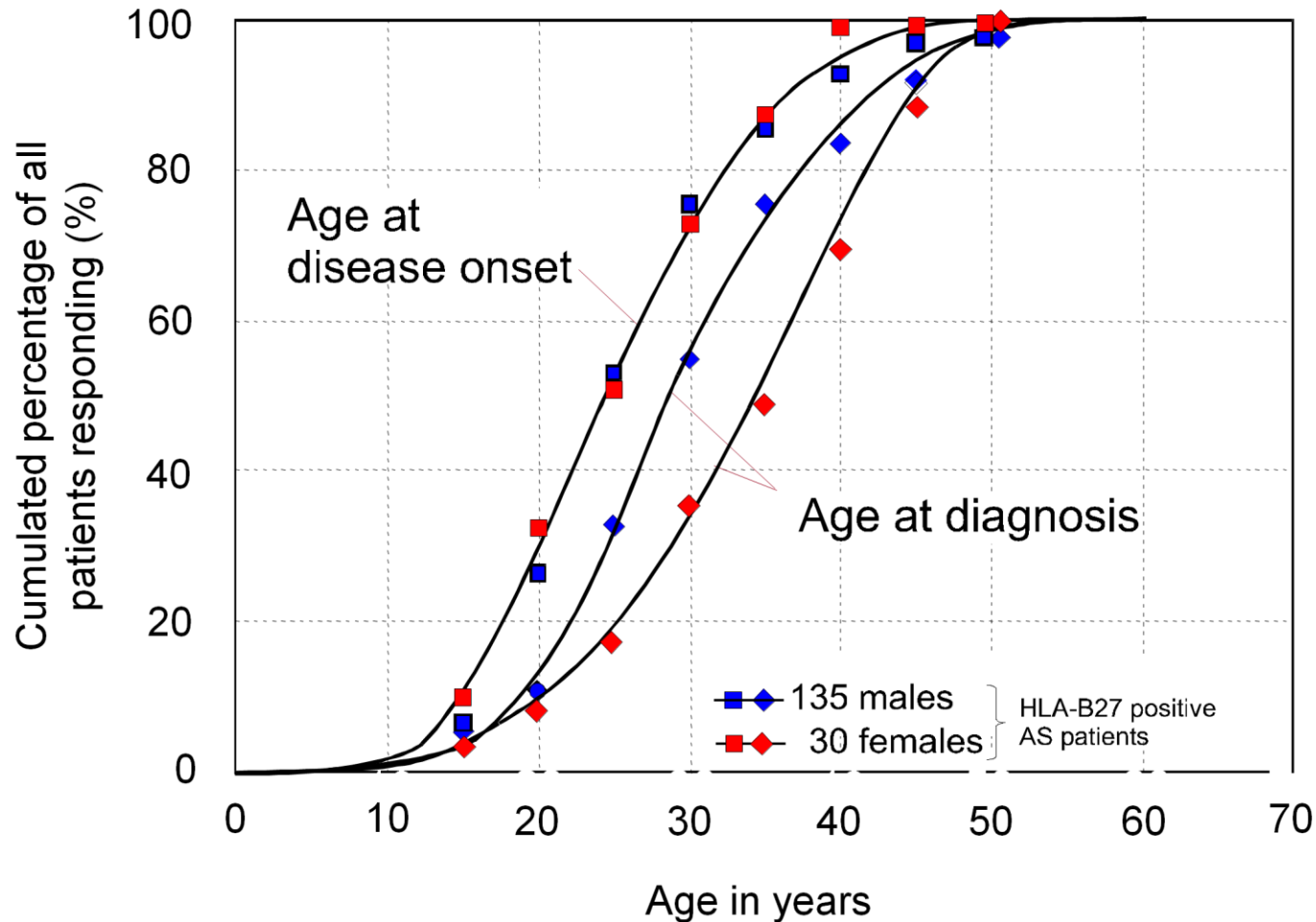
Sensitivity: 79.6%; Specificity: 72.4%

Inflammatory back pain present if at least 4 out of 5 parameters are fulfilled.

Possible Screening Approach for Axial SpA Among Patients with Chronic Low Back Pain



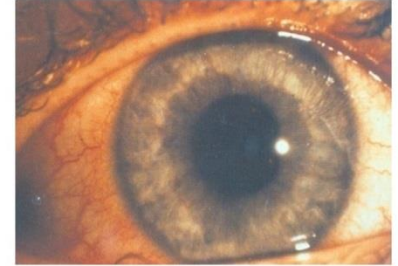
Age at Onset and Time of Ankylosing Spondylitis-Diagnosis



Spondyloarthritis: Characteristic Parameters Used for Diagnosis I

Symptoms

Inflammatory
back pain



Imaging



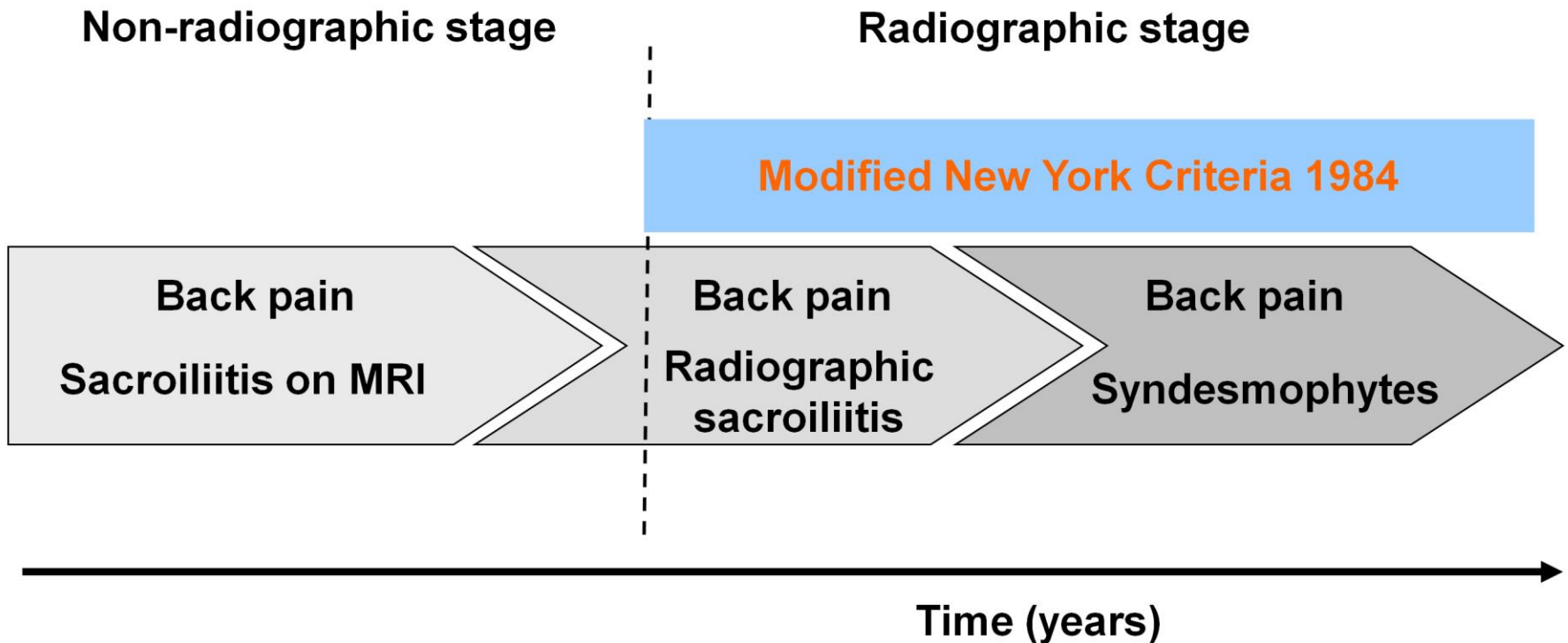
Lab

ESR/CRP

Patient's history

Good response to NSAIDs

Axial Spondyloarthritis



Worker Participation in AS

Comparison with Population

work status

- Employed: 1.1–1.3x lower ($\text{♂} > \text{♀}$)
- Work disabled: 3.0x higher than population

sick leave

- Episode: 15-50%/yr/worker (pop. 6-10%)
- Days: 6-22/yr/worker (pop. 9-12)

presenteeism

- Decrease in productivity of 8% (WLQ)
- 1.9 hours per 2 weeks to compensate

Review of Cost of Illness (COI) Studies

Mean total costs : € 9374/pt/yr

(including work disability)

AS

Weighted mean [IQR]

Direct healthcare and non healthcare costs

Healthcare costs

1992 [1359-2474]

Outpatient costs

1400 [1114-1419]

Inpatients costs

592 [245-983]

Patient & family costs

1104* [541-1432]

Costs of paid productivity loss

Costs sick-leave

913 [388-1079]

Productivity costs (HCA)

6278 [5111-7725]

Productivity costs (FCA)

2271 [1572-2970]

*: includes informal care

Loss of Household Budget for Patients with AS

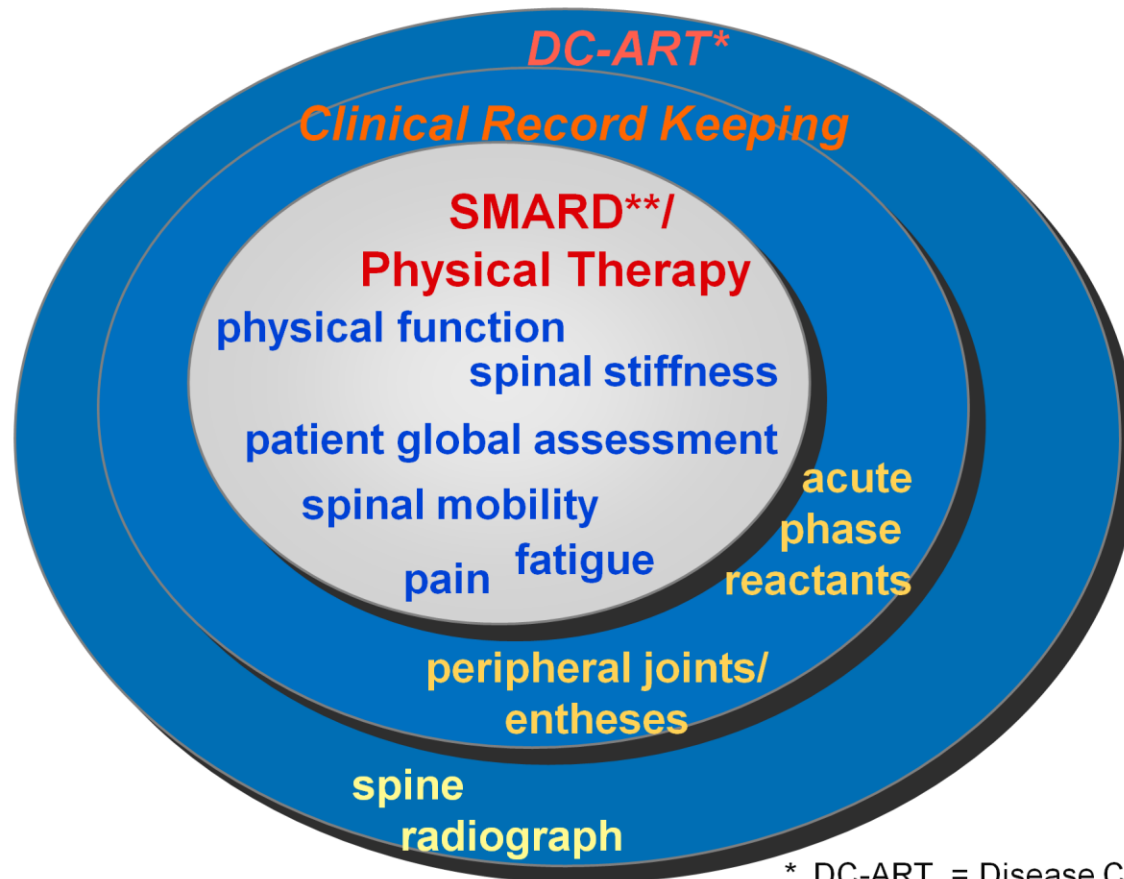
due to out-of-pocket expenditures and income loss

Ankylosing spondylitis

€ 1372^{pt/yr}

AS is an empoverishing disease!

ASAS/OMERACT Core Domains for Ankylosing Spondylitis



- * DC-ART = Disease Controlling-Anti-Rheumatic Treatment
- ** SMARD = Symptom Modifying-Anti-Rheumatic Drugs