

DocSpot – what patients want to know

Professor Chris Denton
Royal Free Hospital
London, UK

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Raynaud's &
Scleroderma
Association

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FAQ



Professor Chris Denton FRCP, Consultant Rheumatologist, answers your questions

Chris Denton is a Professor in the Centre for Rheumatology at the Royal Free Hospital in London and runs the Scleroderma Clinic. He also has a major research programme that spans both clinical and laboratory trials related to scleroderma. He is a medical advisor to the RSA and very grateful to the questions that he is challenged with in our newsletters (Hot News).



Do doctors know the cause of Raynaud's?

Raynaud's phenomenon results from excessive spasm of the blood vessels in the extremities (e.g. fingers, toes, ears etc.). This spasm is triggered by cold or emotional stress, which can be a normal response. It causes symptoms in up to 1 in 10 people, so is very common, but usually not associated with any other disease (primary Raynaud's phenomenon). It probably is caused by complex alterations in the balance of chemicals that narrow blood vessels and those that widen or relax the muscle in the blood vessel wall. Other mechanisms may include an increased sensitivity of nerve endings in the blood vessel wall or damage to the small vessels.

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I have been diagnosed with scleroderma and the biggest problem I have is with itching – why is this and what can be done?

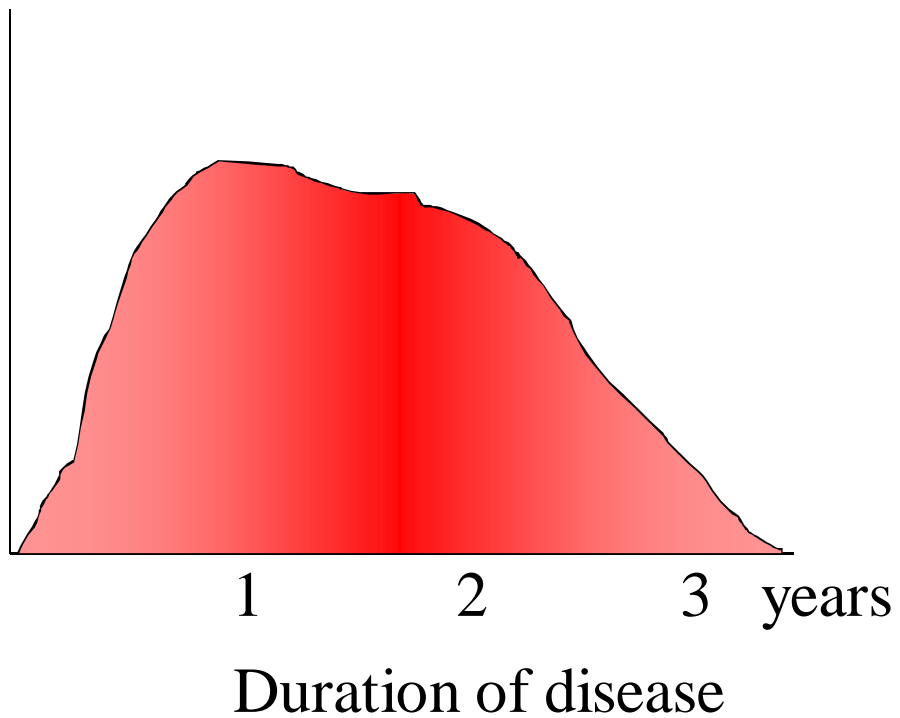
I have been diagnosed with scleroderma and the biggest problem I have is with itching – why is this and what can be done?

- Itching of the skin (pruritis) is common – especially in dcSSc
 - Tends to occur in early disease
 - Due to mast cell degranulation and inflammatory cells and mediators
- Almost always gets better when the skin score falls – improvement is a sign that disease is stabilising
- Other medical causes:
 - Skin disease – eczema, dermatitis herpetiformis (coeliac disease)
 - Iron deficiency, thyroid disease
 - Liver or kidney problems (PBC)

Natural history of itching in scleroderma

- intensity

In diffuse SSc itching is a sign of activity



Management of itching in scleroderma

- Determine any other medical cause
- Skin care
 - emollients/moisturisers/counter-irritants
- Medication
 - antihistamines (drowsiness)
 - corticosteroids– low dose oral prednisolone (topical disappointing)
 - mast cell stabilisation – ketotifen – disappointing results
 - Leukotriene inhibitors – montelukast – anecdotal benefit
- Treat the underlying disease
 - immunosuppression – MTX, MMF etc ...
 - topical MMF
 - topical tacrolimus

**I have Raynaud's and feel tired all the time
and wonder if there is a link between
Raynaud's phenomenon and chronic
fatigue syndrome?**

I have Raynaud's and feel tired all the time and wonder if there is a link between Raynaud's phenomenon and chronic fatigue syndrome?

- There is no direct link
- However any chronic health problem can affect general wellbeing.
- Some associated CTD may cause profound fatigue (SLE, SSc, SS)

Consider the Raynaud's

- Definition of the type of RP
- Try to minimise the impact of Raynaud's on lifestyle
 - Education, drugs, supplements

Treatment of fatigue ...

Medical approach:

- General medical assessment –
 - e.g. thyroid disease (in 30% of SSc)
 - anaemia etc...
 - SSc
 - depression
- Treatment of underlying connective tissue disease if present (e.g. hydroxychloroquine, azathioprine, low dose prednisolone)
- In SLE graded exercise programmes have shown promise in clinical trials

I have hard lumps of “chalk” under my skin and have CREST – what does this mean?



CREST

- **C**alcinosis
- **R**aynaud's
- **oE**sophageal dysmotility
- **S**clerodactyly
- **T**elangiectasia



Major clinical subsets of systemic sclerosis

Limited cutaneous SSc

Diffuse cutaneous SSc



Calcinosis







Mechanism of calcinosis in scleroderma

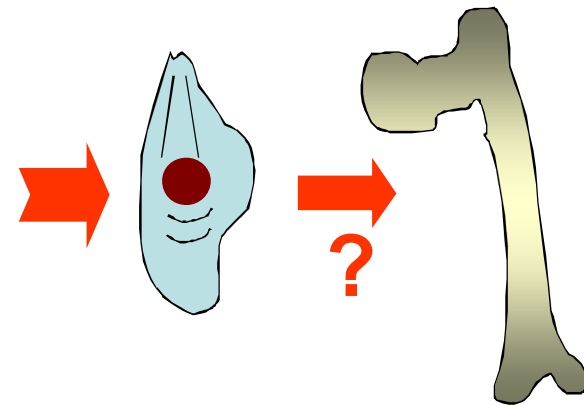
- Calcinosis

- deposition of calcium containing salts in soft connective tissue - calcium hydroxyapatite
- NOT just in CREST – also late stage dcSSc

- Cause:

- Trauma
- Poor blood supply
- Altered extracellular matrix
- Genetics
- Abnormal metabolism - TGF β 1 and BMPs

- **Research ongoing** (Ariane Herrick, Hope Hospital)



Treatment of calcinosis

- Simple approaches:
 - Wax massage
 - avoid interference
 - Treat secondary infection
- Surgical excision
 - open surgery
 - dental drill extraction
 - (Lithotripsy – sound shock waves)
- Medical therapy – none proven
 - optimise Raynaud's therapy
 - Diltiazem? Warfarin?
 - Bisphosphonates
 - Minocycline
 - Anti-TNF α and methotrexate in severe cases?



The opposite of calcinosis – acro-osteolysis



My sister has rheumatoid arthritis – is there any association with me having scleroderma?

Overlap syndromes

Systemic sclerosis (SSc) is associated with clinical and serological features of another connective tissue disease (inflammatory arthritis, myositis, Sjögren's syndrome, lupus).

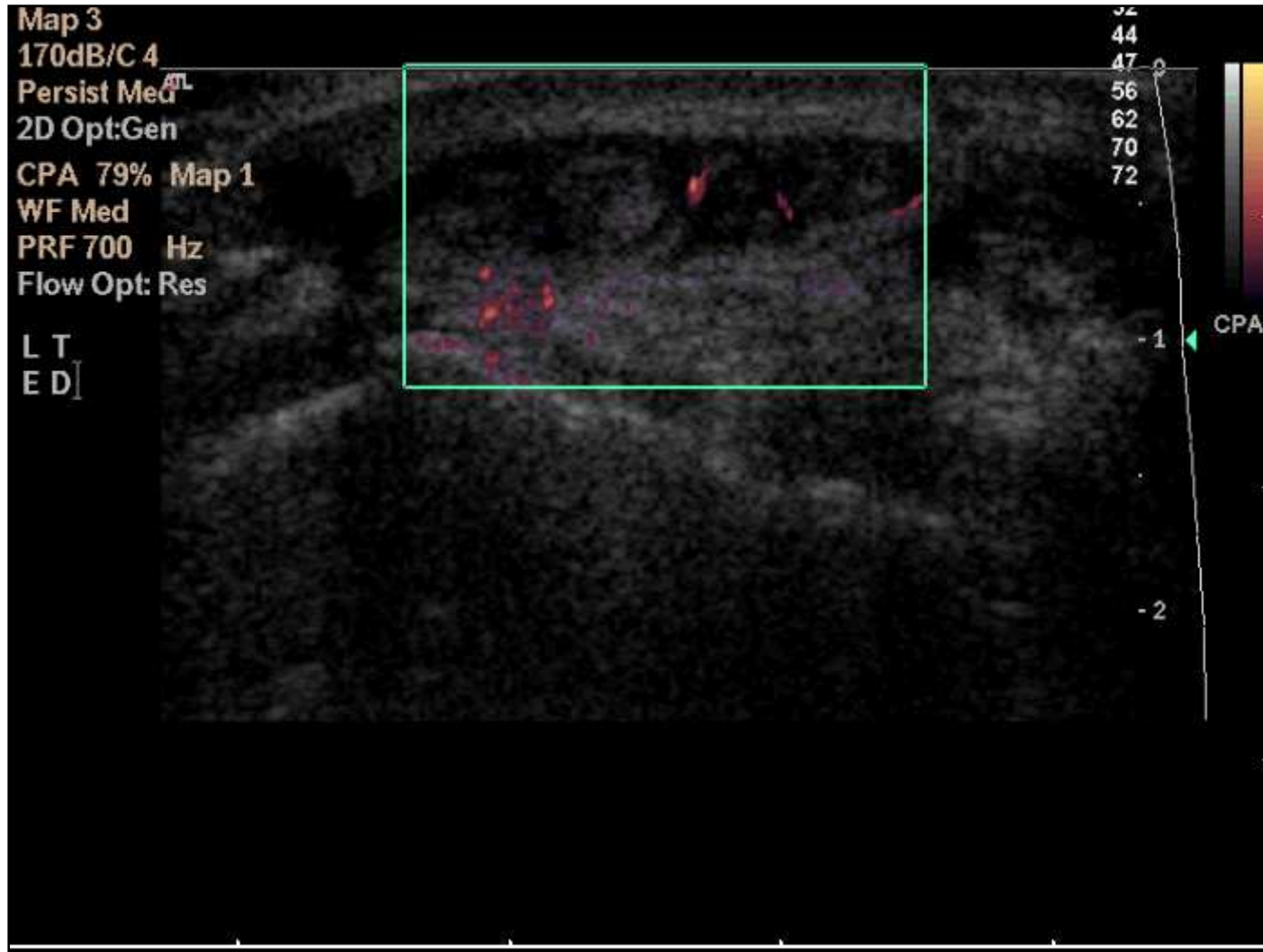


RA-scleroderma overlap syndrome

- RF positive
- arthritis - may be erosive
- scleroderma - ANA positive
- often SSc-specific serology



Power doppler USS hands



Extensor digitorum tenosynovitis

Frequency of overlap SSc in the Royal Free Hospital scleroderma cohort

The Journal of
Rheumatology

The Journal of Rheumatology

Volume 38, no. 11

Clinical and Serological Hallmarks of Systemic Sclerosis Overlap Syndromes

ANGELA PAKOZDI, SVETLANA NIHTYANOVA, PIA MOINZADEH, VOON H. ONG,
CAROL M. BLACK and CHRISTOPHER P. DENTON

J Rheumatol 2011;38;2406-2409

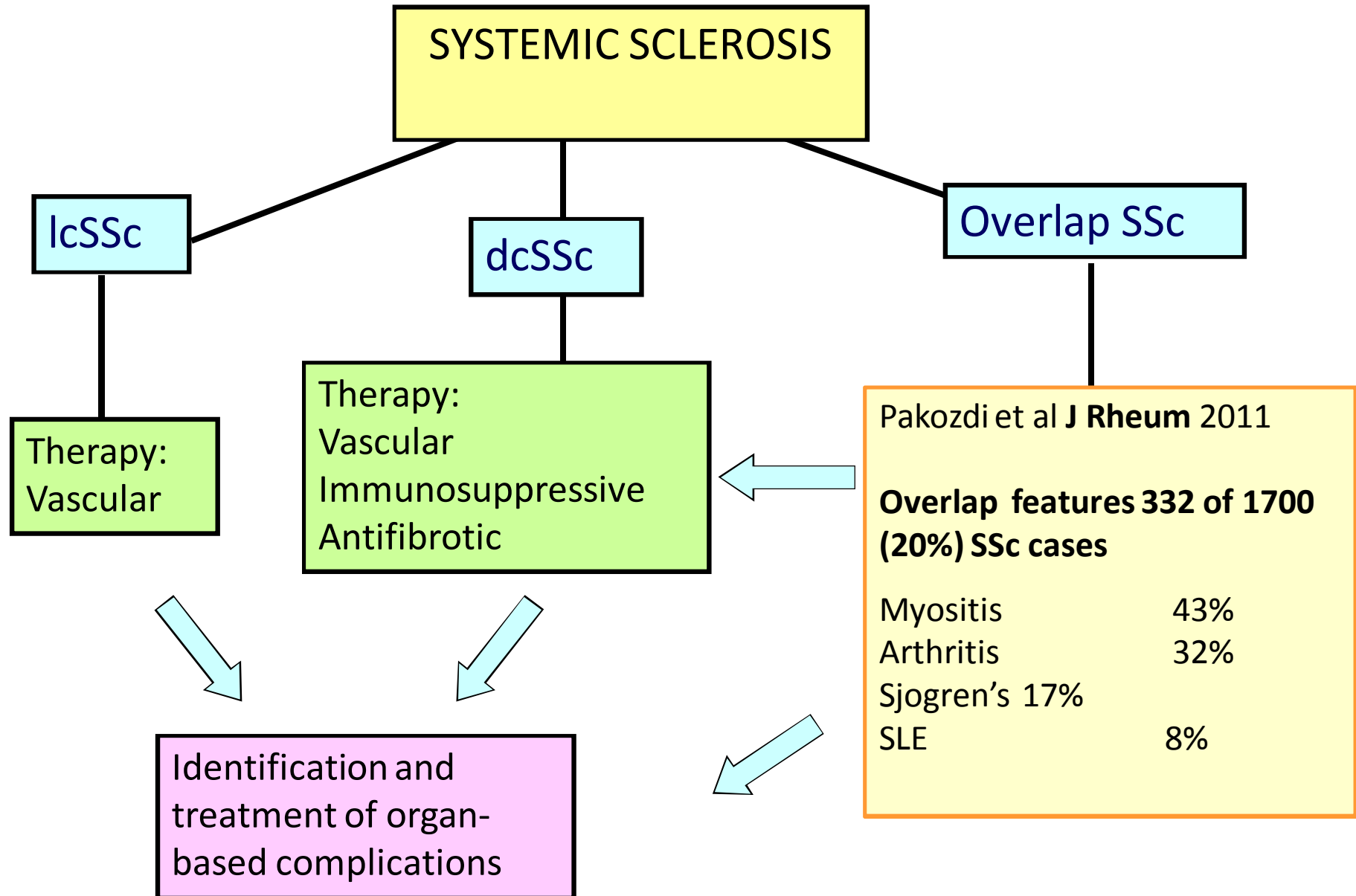
<http://www.jrheum.org/content/38/11/2406>

Pakozdi et al, **J Rheum** 2011; **38**:2406-09

Distribution of patients with SSc overlap syndromes

All overlap SSc	339	(100%)
SSc/myositis	128	(37%)
SSc/inflammatory arthritis	95	(28%)
SSc/Sjögren syndrome	52	(15%)
SSc/lupus	27	(8%)
≥ 3 CTD	37	(12%)

Management of systemic sclerosis



Why do I have so much pain in
my hands?

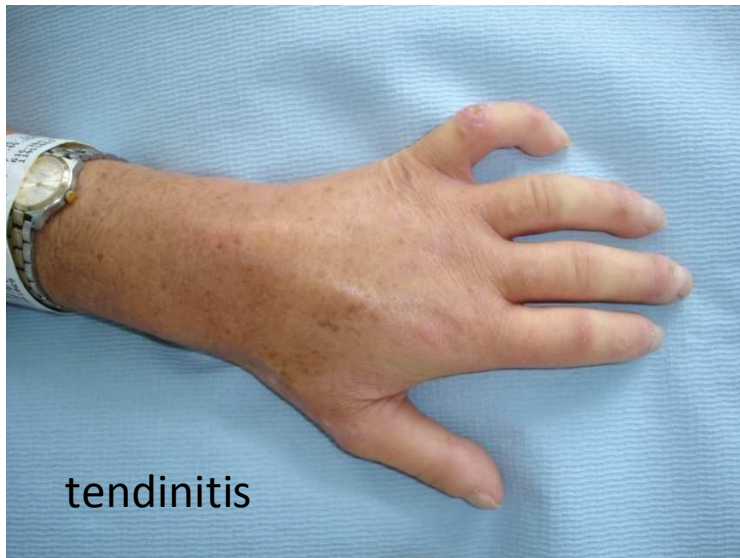
Causes for musculoskeletal pain in systemic sclerosis

- Contractural arthropathy
 - Contractural tendinopathy
 - Arthritis
 - Myositis
 - Fasciitis
 - Connective tissue atrophy
 - Calcinosis
 - Neuropathic pain
 - Ischaemic digital vasculopathy
- Sclerotic
- Inflammatory
- Dystrophic
- Neurovascular

Common musculoskeletal manifestations of systemic sclerosis



Rheumatoid arthritis overlap



tendinitis



arthritis

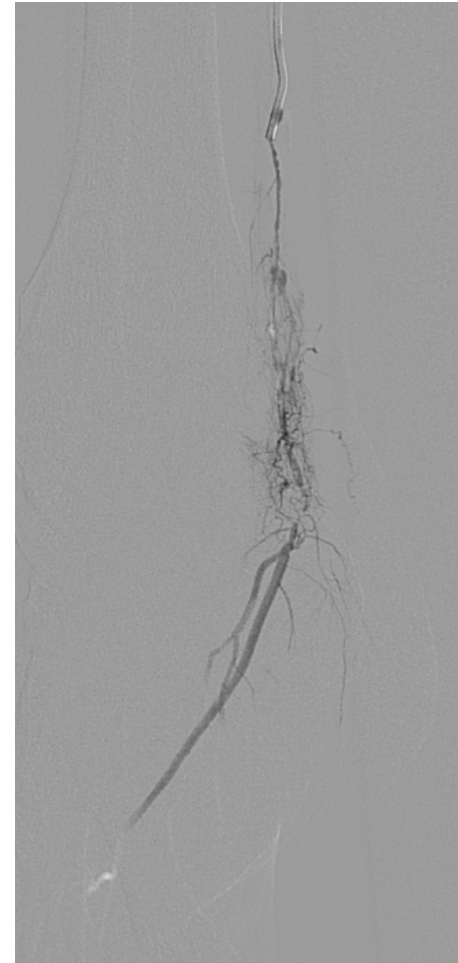
Other causes for musculoskeletal pain in systemic sclerosis



Massive calcific peri-arthritis



Gangrene associated with macrovascular occlusion



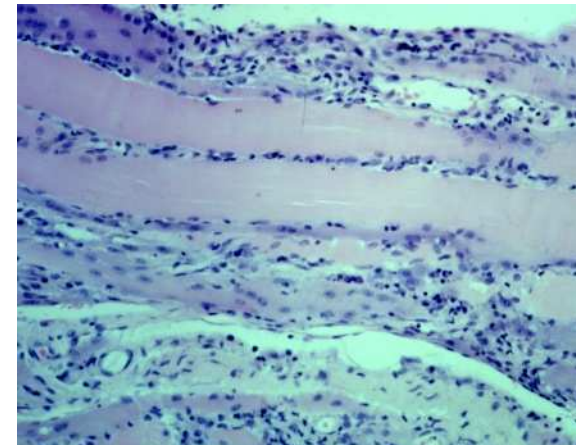
Therapy for upper limb pain in scleroderma

- Pain killers
- Physical therapy
- Disease modifying drugs for arthritis (DMARDs)
- SSc management
- Multidisciplinary management
 - Pain
 - Vascular specialists
 - Nurses
 - Psychologist
 - Physician
 - Surgeon
 - Physiotherapist
 - Occupational therapist

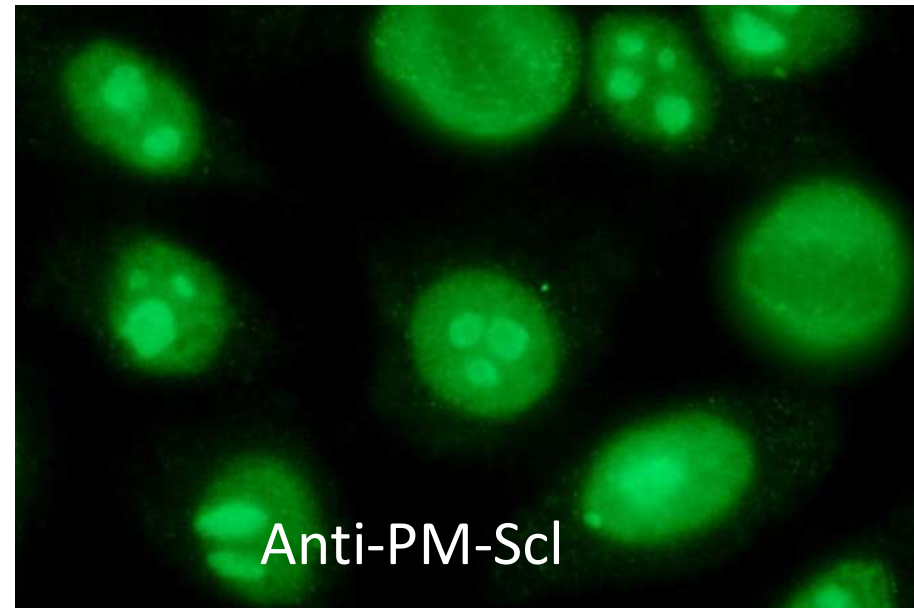
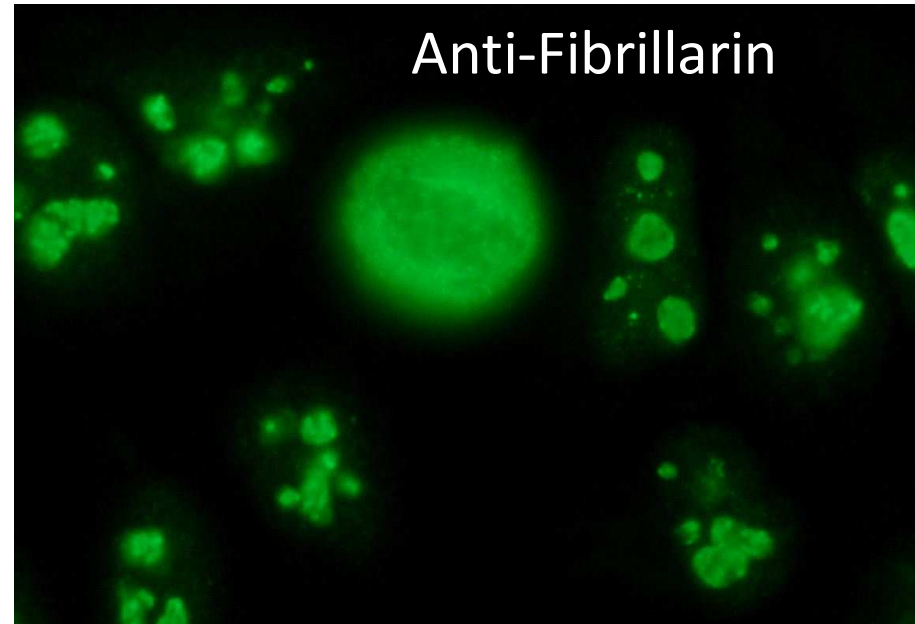
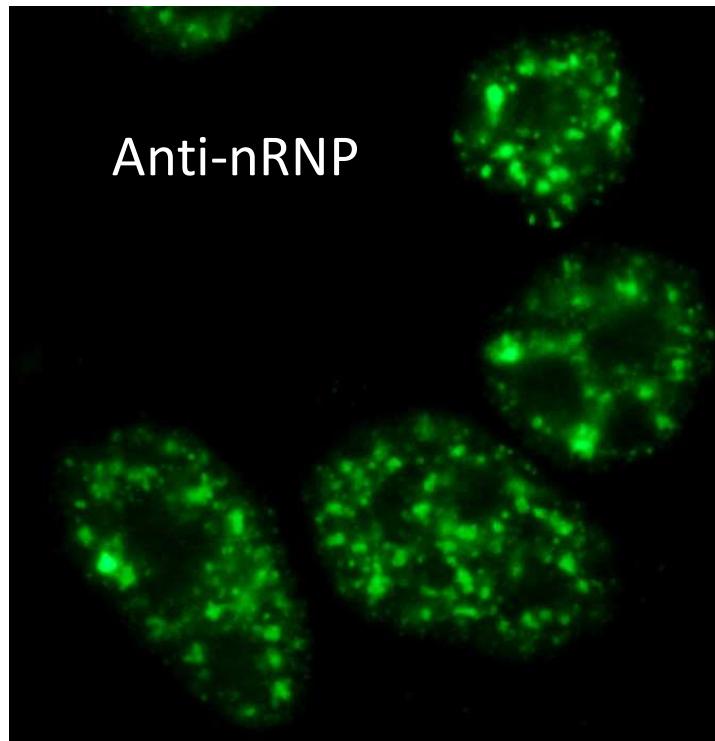
I have scleroderma and cannot get out of a chair unaided – why?

SSc/myositis overlap

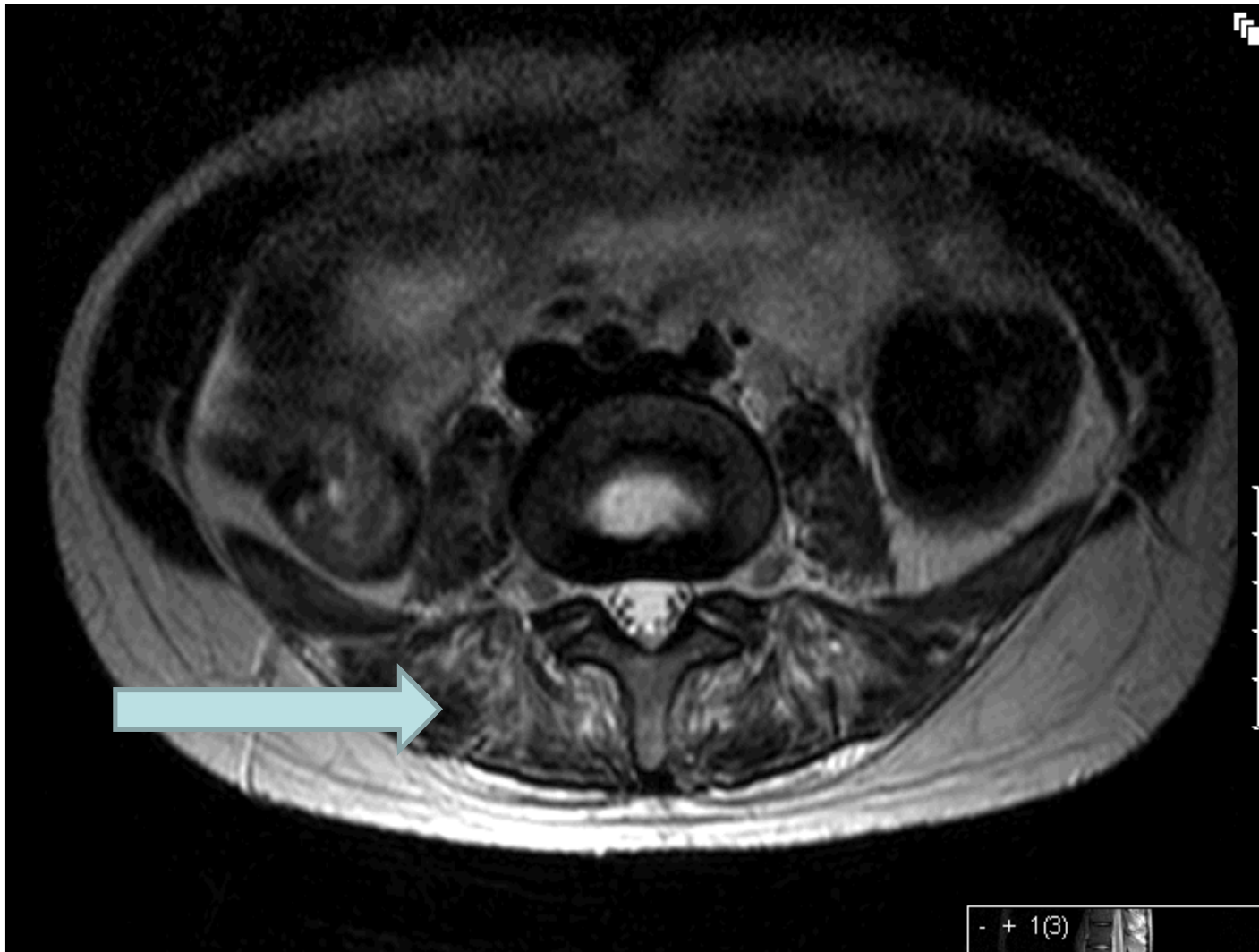
- The prevalence of the diffuse subset in SSc/myositis overlap is relatively high (40%) compared to other SSc overlap syndromes (17-26%).
- PM-Scl is the main autoantigen in patients with SSc/myositis overlap syndrome
- nRNP and U3RNP also associated frequently found in SSc/myositis overlap



Scleroderma hallmark
autoantibodies
associated with overlap
myositis



MRI Lumbar spine – back pain and weakness



T2W image

Why am I taking drugs for arthritis
when I have been diagnosed with
scleroderma?

Immunosuppressive therapy for dcSSc

Potential immunomodulatory strategies

Methotrexate

Cyclophosphamide

Azathioprine

Mycophenolate mofetil

Anti-thymocyte globulin

Ciclosporin, Tacrolimus,

Basiliximab, Rituximab

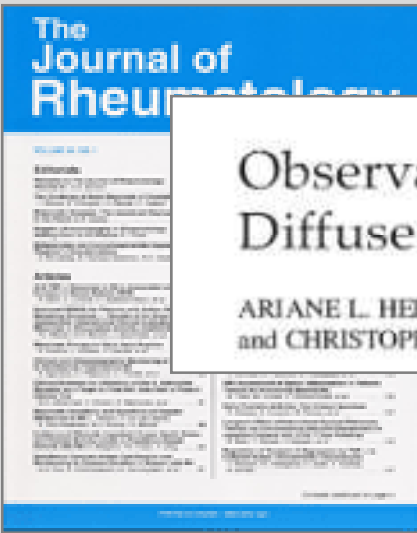
Rapamycin

Stem cell transplant (autologous/allogeneic)

Oral tolerance to type I collagen

Early stage dcSSc

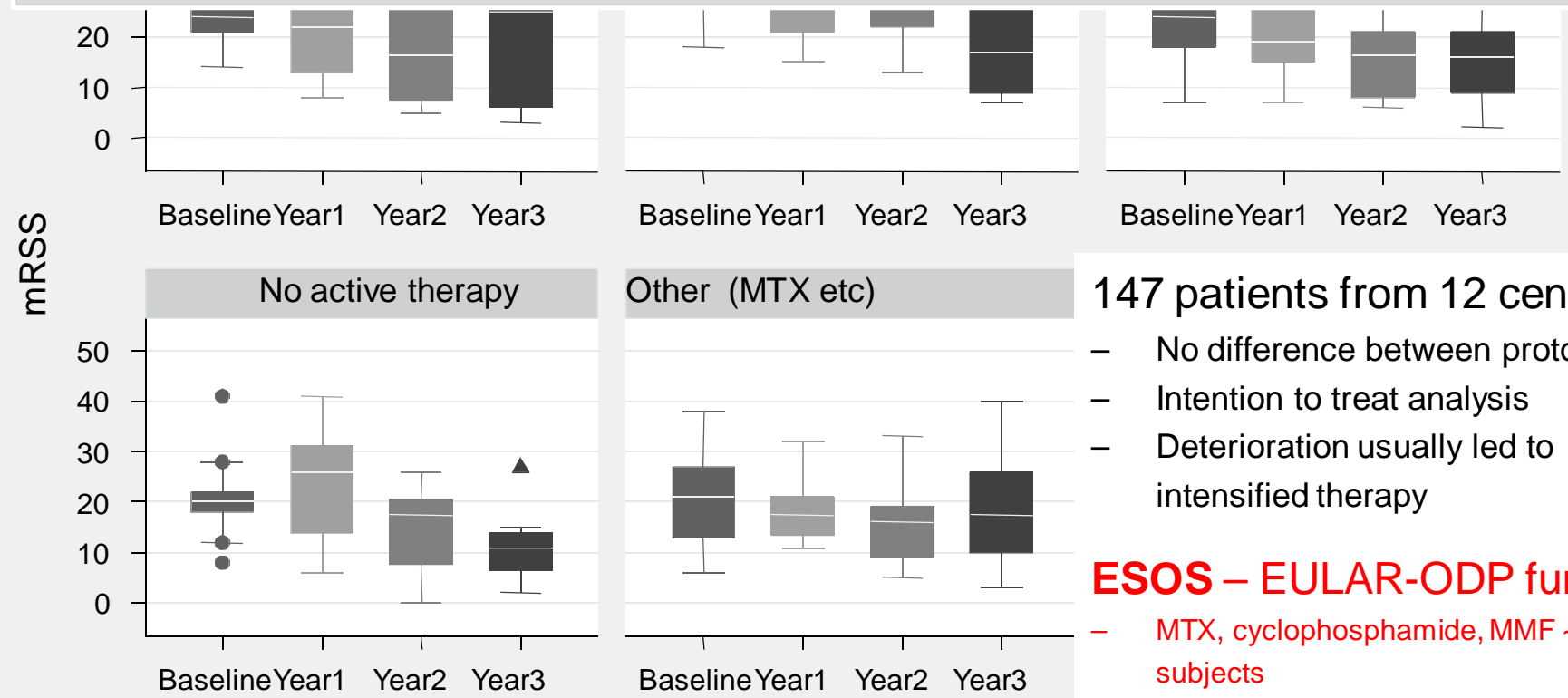
Late stage dcSSc



Observational Study of Treatment Outcome in Early Diffuse Cutaneous Systemic Sclerosis

ARIANE L. HERRICK, MARK LUNT, NINA WHIDBY, HOLLY ENNIS, ALAN SILMAN, NEIL McHUGH, and CHRISTOPHER P. DENTON

Herrick et al, *J Rheum* (2010); **37**: 116-24.



147 patients from 12 centres

- No difference between protocols
- Intention to treat analysis
- Deterioration usually led to intensified therapy

ESOS – EULAR-ODP funded

- MTX, cyclophosphamide, MMF ~400 subjects

Herrick et al, *J Rheum* (2010); **37**: 116-24.

Any more questions?

