Myositis:
Getting in Sync with your Healthcare team

Namita Goyal, MD
Associate Professor of Neurology
Associate Director, Neuromuscular Center
UC Irvine

The Myositis Association
Kentucky
September 7, 2018

UC Irvine Health
School of Medicine

Overview of Care Team: Checklist

- Rheumatologist/Neurologist
- Primary Care Physician (PCP)
- Pulmonologist/Respiratory therapist
- Cardiologist
- Speech/Swallow therapist
- Physical therapist/Occupational therapist
- Dietician/Nutritionist
- Social Worker

Neurologist/Rheumatologist:

Initial Evaluation:

- History/exam
- Diagnostic evaluation
  - Blood tests: CK levels
  - Antibodies
  - EMG
  - Muscle biopsy
  - Muscle MRI
- Cancer Screening in Dermatomyositis
  - Esp if >40 yrs old
  - up to 3-5 years from symptom onset

Myositis Antibodies

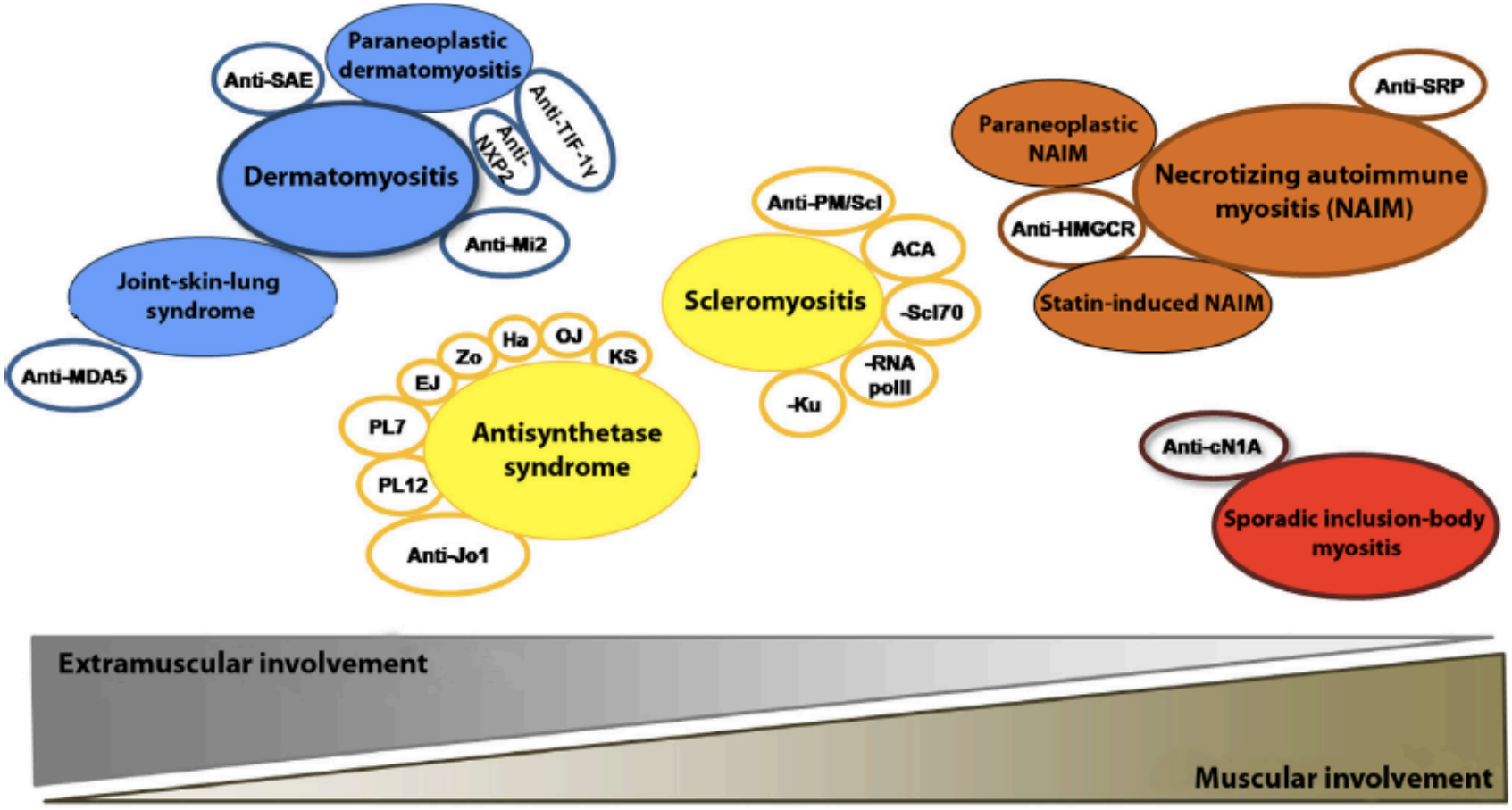


Fig. 2. Clinical and serological diversity of inflammatory myopathies (IM). The prominence of the extramuscular manifestations is usually in inverse proportion to the severity of the muscle involvement. The autoantibody profile can be used to identify patient subgroups with globally homogeneous clinical features and outcomes.

UC Irvine Health
School of Medicine

A. Meyer et al. / Joint Bone Spine 85 (2018) 23–33

Myositis Specific Antibodies: Dermatomyositis Autoantibodies

Mi-2	classic cutaneous manifestations, **respond well to immunotherapy**, low risk of cancer
TIF1γ	**high risk of malignancy**, classic skin manifestations, "diffuse photoerythema, dusky red face"
NXP-2	subcutaneous **calcifications**, (in up to 25% of juvenile DM, but also adults), increased risk of **malignancy**
MDA5	**rapidly progressive ILD**, (20-30% of Asian DM patients, less freq in Caucasians), **skin ulcerations**, tender palmar papules, oral ulcers, **minimal muscle** involvement (clinically amyopathic)
SAE	least frequent, <10%, dysphagia, skin disease, good prognosis

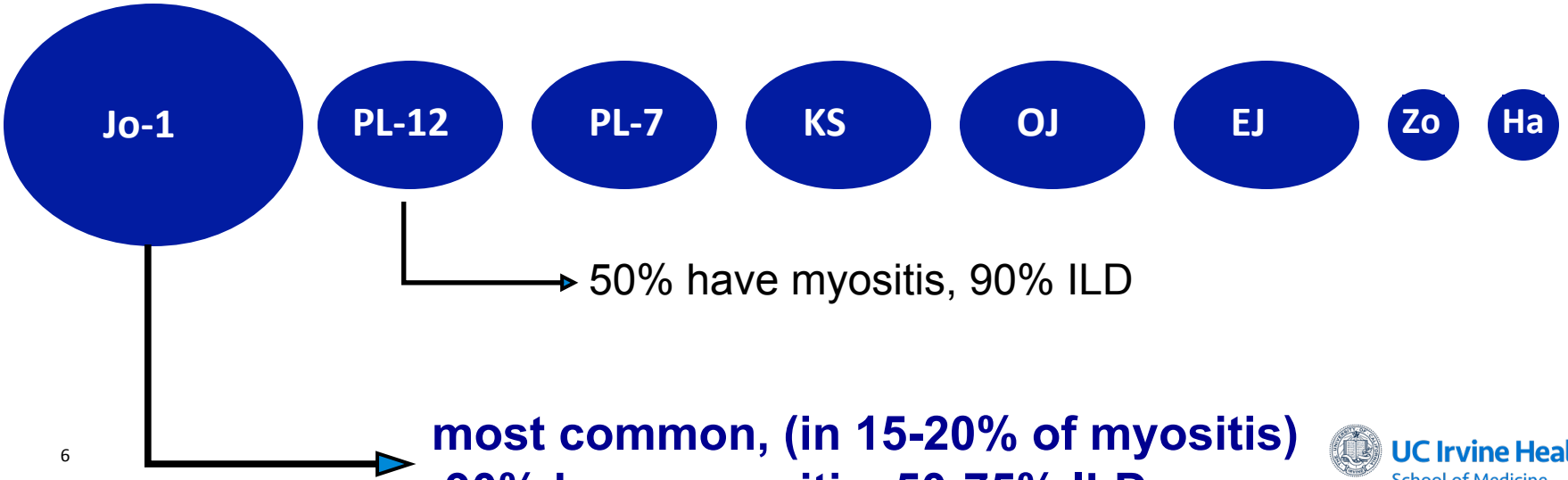
UC Irvine Health
School of Medicine

Myositis Specific Antibodies (MSA):
Antisynthetase Autoantibodies

Antisynthetase syndrome:

- Myositis, Interstitial lung disease (ILD), inflammatory arthritis, fever, Raynaud's phenomenon, mechanic's hands
- Some have prominent skin rash

8 Abs, most common MSA, identified in 35-40% of myositis



UC Irvine Health
School of Medicine

Myositis Associated Antibodies (MAA):

Nonspecific, in myositis & connective tissue diseases

- Ro52/TRIM21, PMScl, ribonucleoprotein complex (RNP; U1 RNP, U2 RNP, U4/U6, RNP, U5 RNP), Ku

Ro52 → **most common**, associated with ILD

PMScl → seen in PM, systemic sclerosis (SSc), & PM/SSc overlap syndrome, associated with lung and esophageal involvement

Ku → in Overlap syndrome, frequent joint involvement, Raynaud's and ILD

UC Irvine Health
School of Medicine

MSA: Immune-Mediated Necrotizing Abs

Prominent myofiber necrosis with minimal inflammation

CK > 1,000-10,000

Anti-SRP

Rare,
(<5% of all myositis)

- Rapidly progressive onset
- Very high CK levels
- Dysphagia
- Neck extensors
- Severe weakness
- May not respond well to immunotherapy

Anti-HMGCR

(6-9% of all myositis)

- First described in context of statin exposure (2010)
- Also in statin-naïve
- (Not found in self-limited statin intolerance)
- May require aggressive immunotherapy or IVIg

Neurologist/Rheumatologist: (follow-up)

Subsequent visits:

- History/exam
- Medication management
- Response to immunotherapy?
  - Adjust medications
  - Lack of response
  - Alternative therapies/clinical trials?
  - Wrong diagnosis?
- Adverse effects of medications
  - Check blood counts, liver, kidney

Primary Care Physician:

- Age appropriate health screening
- Monitor Blood sugars, Blood pressure – if on steroids
- Bone density test (Vitamin D with Calcium)
- Check in with PCP if not feeling well, may not mount fever if immunosuppressed
- Vaccinations
- Help with Mood/Antidepressant?

Pulmonologist/Respiratory therapist:

- If Interstitial lung disease (ILD)
  - CT Chest (to screen and monitor progression)
  - Pulmonary function tests

- Noninvasive Ventilation (BiPAP)
  - Quite beneficial in respiratory insufficiency
  - Difficulty tolerating?
    - Work with respiratory therapist to adjust mask/settings

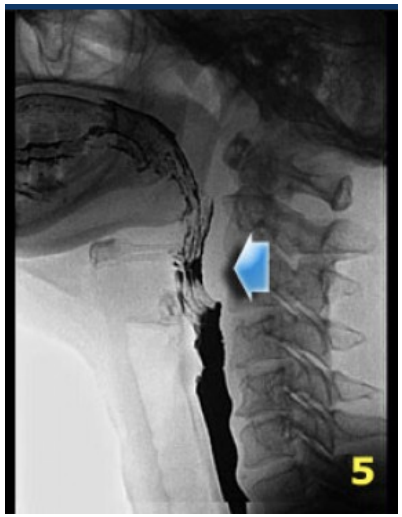
UC Irvine Health
School of Medicine

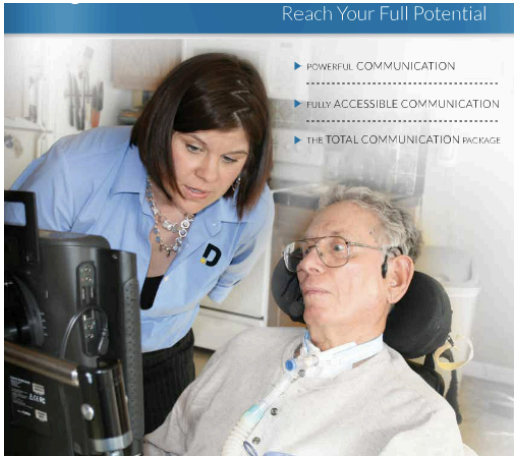
Cardiologist:

- Cardiomyopathy/cardiac arrhythmias
- Rare, but potential complication of DM, Anti-synthetase syndromes
- Close monitoring
- Medical management
- Echo/EKG

Speech/Swallow Therapist:

- Difficulty swallowing/dysphagia
  - Can be leading cause of morbidity/ mortality
  - Up to 1/3 of myositis patients (esp IBM)
- Barium Swallowing evaluation
  - Can detect subclinical involvement
  - Evaluates severity
- Modified diet
- If severe, G-tube
  - Reduces risk of aspiration pneumonia
- Communication devices





Physical/Occupational therapists:

- Risk of falls
- Adaptive equipment needs
  - Foot brace (AFO), Cane, Walker, Scooter, Wheelchair
  - Shower chair
  - Hospital bed
- Home safety evaluation – railings, grab bars
- Role of exercise
  - No pain, no gain- NOT the motto!
  - Stationary cycling, pool therapy (if safe to get in)
  - Don't exercise to the point of pain or significant fatigue

Dietician/Nutritionist:

- Well-balanced diet
- Maintaining weight
- If on steroids, weight gain is a recognized concern
  - Ask for a consult with nutritionist
- If difficulty swallowing, weight loss is a concern
- With muscle atrophy, weight loss may occur
- G-tube (when severe dysphagia) – reduces risk of aspiration
  - Maintain calories and weight
  - Helpful for pills
  - Hydration
- Constipation (common), esp in impaired mobility

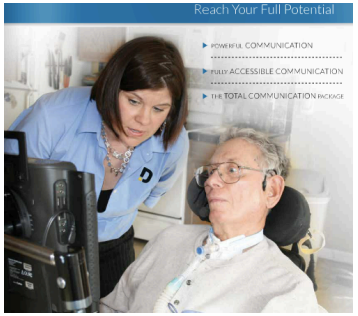
Social Worker:

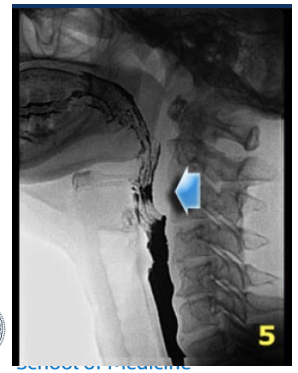
- Care giving resources
- Home health services
- Support groups
- Psychologists/psychotherapists

Other Specialists:

- Dermatologist – dermatomyositis patients with severe skin involvement
- Gastroenterologist (GI) – if PCP has difficulty managing and more severe GI issues
- Psychiatrist

Goal: Multidisciplinary Team Approach





Neurologist/ Rheumat

Respiratory therapist

Physical/ Occupational Therapist

Swallow Therapist

Speech Therapist

Social Worker

Dietician

Medical assistant/ Nurse

Other specialists

Patient