Myositis: 
Getting in Sync with your Healthcare team

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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.

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
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

Jo-1  PL-12  PL-7  KS  OJ  EJ  Zo  Ha

50% have myositis, 90% ILD

most common, (in 15-20% of myositis)
90% have myositis, 50-75% ILD
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
**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
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
- Other specialists
- Medical assistant/Nurse
- Dietician
- Swallow Therapist
- Social Worker
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