Naval Medical Center Portsmouth
Rheumatology Referral Guidelines

Diagnosis:	**Elevated Creatine Kinase (CK) and Idiopathic Inflammatory Myopathies (IIM)**

Clinic Name	Rheumatology
Clinic Phone Number	757-952-2160 or 2161
On Call Numbers	757-860-5702

1. Indications for Specialty Care:

- No prior Rheumatology evaluation and inflammatory myositis (IIM) is suspected based on:
  - Insidious onset of proximal muscle weakness (shoulders and hips), elevated muscle enzymes, with supportive EMG findings, muscle inflammation on MRI, or typical skin manifestations (Dermatomyositis, DM).
  - Other mimics of myositis have been excluded.

- When specific organic pathology is present (evidence of end organ damage or criteria present for systemic rheumatic illness such as SLE, RA, scleroderma, etc.).

*****Referral should not be given to merely reaffirm a previous diagnosis of a non-rheumatic disease causing increased CK or non-autoimmune muscle disease. *****

2. Quality Consult Criteria

When referring a patient, please include as much of the following information as possible (OK to cut and paste this into consult request)

1. Provisional diagnosis
2. Duration of Problem
3. Prior treatments
4. Current treatments/medications
5. Diagnostic studies obtained (imaging, labs, other tests, etc.)
6. Primary reason for consult
7. Use of referral guidelines

3. Diagnosis Definitions

- The idiopathic inflammatory myopathies (IIM), including polymyositis (PM), dermatomyositis (DM), and inclusion body myositis (IBM), are primary autoimmune causes of muscle disease, and secondary myositis may also be seen in established connective tissues diseases (e.g. Lupus).

- Muscle damage and elevated muscle enzymes (e.g. CK) are relatively common in clinical practice. Numerous disorders may result in muscle injury, including primary metabolic myopathies, endocrine myopathies, nutritional defects, electrolyte disorders, muscular dystrophies, as well as neuromuscular disorders (e.g. ALS). Infections, medications (e.g. statins) and drugs (e.g. alcohol) are common culprits.

4. Initial Diagnosis and Management

- The diagnosis of myositis is an affirmative diagnosis made clinically in accordance with 2017 ACR / EULAR classification criteria. **IIM is a diagnosis of exclusion**, and thus history, physical exam findings, laboratory tests and imaging studies play an important role in establishing the diagnosis according to the ACR's criteria.

- Classification of IIM is best summarized in: 2017 European League against Rheumatism/American College of Rheumatology Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and Their Major Subgroups Arthritis & Rheumatology, Vol. 69, No. 12, December 2017, pp 2271–2282.

- A thorough history is essential in uncovering the underlying etiology of muscle damage.
  - A history of trauma, crush-injury, recent surgery, or seizures should be elicited.
  - Medication usage (including statins, niacin, fibrates, AZT, diuretics, laxatives, colchicine, vincristine, hydroxychloroquine, chloroquine, amiodarone, neuroleptics) may indicate a medication-induced myopathy.
  - Significant alcohol or illicit drug usage (e.g. cocaine) may indicate toxin-induced myopathy.
  - Exercise-induced weakness or rhabdomyolysis, muscle pain/cramping, 2nd wind phenomenon or precipitating factors such as illness/fasting/cold exposure may indicate a metabolic myopathy (glycogen/lipid/mitochondrial disorders).
  - A family history of muscle disease may indicate a muscular dystrophy (e.g. Limb-girdle, fascioscapulohumeral). Neuromuscular disease should also be considered (e.g. ALS).
  - Clinical features suggestive of endocrine disorders including Cushing's, Addison's, thyroid disease, hyperparathyroidism, and diabetes should be elicited.
  - A history of preceding infections - bacterial (e.g. staph), parasitic (e.g. toxoplasma, trichinella) or viral infection (e.g. influenza, EBV, CMV, HIV, HSV, adenovirus, etc.) - should be sought.
  - History of systemic symptoms (fevers, weight loss, fatigue, anorexia), rashes or subcutaneous nodules, joint pain/swelling or prolonged AM stiffness, dysphagia, dysphonia, Raynaud's, shortness of breath, or history of cancer may indicate an autoimmune process.

- A full general physical exam is essential with attention to pattern of muscle weakness / symmetry, inflammatory arthritis, rashes/subcutaneous nodules, findings of interstitial lung disease (e.g. crackles), and Raynaud's phenomenon.

- Lab evaluation is helpful, but rarely definitive in evaluating rheumatic complaints. Markers of muscle damage include CK, aldolase, AST and ALT, LDH, myoglobin**. THERE IS NO SUCH THING AS A RHEUMATOLOGY PANEL**. ANA can be negative in IIM, and inflammatory markers are neither sensitive nor specific. Labs to assess for renal/liver disease, electrolyte abnormalities (Ca, Mg, Phos, K), resting lactate, and any suspected endocrinopathies (e.g. TSH) may be helpful. Myositis specific antibodies may be helpful in confirming a strong clinical suspicion of IIM.

- EMG / nerve conduction studies help differentiate neuropathic from myopathic processes.

- Imaging modalities such as MRI (e.g. thighs) may help identify inflamed muscle and target biopsies.

- Features suggestive of PM or DM include insidious onset of symmetric proximal muscle weakness, elevated muscle

enzymes, with myopathic changes on EMG and symmetric muscle inflammation on MRI. Typical skin manifestations of DM include heliotrope rash (around eyes), Gottron's papules (across knuckles), and abnormal nailfold capillaries with Raynaud's phenomenon. Skin biopsy in DM shows interface dermatitis.

- Features suggestive of IBM include age >50 years, proximal and distal muscle weakness (e.g. finger flexors), mixed myopathic/neuropathic EMG, with a poor response to treatment (e.g. steroids).

5. Ongoing Management and Objectives

- While the evaluation is underway, management in clinically stable patients is usually supportive.
  - Supportive care with physical and occupational therapy is essential to maintaining functionality for many different types of myopathy.
- Potentially culprit medications should be held, and the muscle enzyme trend should be monitored.
- Routine cancer screening should be updated.
- A neuromuscular evaluation with EMG / nerve conduction testing and MRI may be very helpful in differentiating potential causes of muscle damage.
- The patient should be referred for muscle biopsy or skin biopsy (of rash) if clinically indicated.
- If IIM is strongly suspected, early referral to a rheumatologist is indicated for initiation of aggressive immunomodulatory therapy.

6. Criteria for Return to Primary Care

- Diagnosis of elevated CK or myopathy established in the absence of autoimmune disease.
- Management questions do not require a re-referral and are preferably handled as a conversation (potentially electronically) between the referring provider and the consulting Rheumatologist.
- Patients with concomitant autoimmune disease and elevated muscle enzymes may be co-managed with Rheumatology.

Date Adopted or Last Reviewed:	01 February 2018	By	CDR Shauna O'Sullivan LCDR Jeffrey Eickhoff LCDR Terrence Kilfoil LCDR Jason Weiner
Referral Guidelines require review every three years.			

7. Resources/References

- 2017 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and Their Major Subgroups Arthritis & Rheumatology, Vol. 69, No. 12, December 2017, pp 2271–2282.
- American College of Rheumatology: https://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Inflammatory-Myopathies