Clinically Amyopathic Dermatomyositis (CADM) In Focus

- Amyopathic Dermatomyositis (ADM) and Hypomyopathic Dermatomyositis (HDM), together referred to as Clinically Amyopathic Dermatomyositis (CADM), are unique subsets of Dermatomyositis (DM), one of the Idiopathic Inflammatory Myopathies, and are both rare, systemic, autoimmune diseases of the skin, with little-to-no muscle involvement typical of "classic" DM, of which the cause is unclear and there is no cure. ***See Dermatomyositis (DM) for more***
- These are also known as "dermatomyositis siné myositis" and "skin predominant dermatomyositis."
- With ADM, there is typically no muscle weakness on exam, nor are there muscle findings on testing. With Hypomyopathic DM, there is no muscle weakness on exam, however patients may have findings of muscle involvement in labs, such as elevated muscle enzymes, and on EMG and MRI. The diagnosis may later change to "classic" DM if muscle involvement becomes more defined.
- CADM affects more women than men and is more common in younger Caucasian and Asian females.
- Skin findings of CADM may include rashes that may go unnoticed with darker skin tones, violet (heliotrope) rash on the face, neck, forearms, upper chest (V-sign across the front of the chest and Shawl sign across the shoulders and upper back) and other areas of the body. Gottron's Papules or Gottron's Sign, mechanic's hands, heliotrope rash on eyelids, with or without swelling, ulcers, calcinosis, and secondary changes, poikiloderma. Intense itching and photosensitivity are common, as is scalp inflammation and thinning of the hair. ***See Dermatomyositis and the Skin for more***
- Risk of an associated cancer is higher in those with CADM for 5 years pre-and-post disease onset and CADM can be present as a part of Mixed Connective Tissue Disease.
- Other symptoms of CADM may include debilitating fatigue, joint pain, weight loss, fevers, trouble swallowing, Raynaud's phenomenon, nonerosive inflammatory polyarthritis, and muscle pain. CADM may affect other organs of the body such as the heart and lungs.
- Diagnosing CADM can be difficult and can often take months or years. A misdiagnosis of Lupus SLE is common. Patient history, skin exam, Myositis-Specific antibodies (MSA), Myositis-Associated antibodies (MAA), other blood tests (muscle enzymes, inflammatory, autoimmune, and cancer markers, and other antibodies), MRI, EMG, nerve conduction studies, and cancer screenings, along with skin and muscle biopsy can assist in making a diagnosis.
- Treating CADM includes sun avoidance, ample use of sunscreen, UVA and physical sun blockers, oral and topical steroids, non-steroidal topicals, and antimalarial agents.
- Off-label use of medications may be used successfully in treating CADM including immunosuppressive agents, chemotherapy medications, and infusion therapies such as immunoglobulin (Ig) and monoclonal antibodies (rituximab). These may cause harsh side effects and a weakened immune system.
- CADM patients with lung involvement such as Interstitial Lung Disease may require the use of oxygen.

Patients with CADM may be covered in itchy and uncomfortable pink-red-violet skin rashes while others may not look sick (invisible illness). For example, patients may have associated internal complications such as cancer, heart or lung disease that are not obvious. You may see some wearing long-sleeves, hats, and gloves, even in hot weather, to protect them from the sun. Each patient is very different.

With all the difficulties in the diagnostic process and living with this disease, the journey takes a toll on patients and families alike. Look closer and put #MyositisInFocus and learn more about Clinically Amyopathic DM at www.UnderstandingMyositis.org/cadm