

# Teaching Slide Collection of the Cutaneous Manifestation of the Myositis Damage Index (MDI)

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# Introduction

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- This collection demonstrates representative examples of the cutaneous lesions associated with juvenile and adult dermatomyositis (DM) and other idiopathic inflammatory myopathies which are included in the Myositis Damage Index.
- Damage is defined as persistent changes in anatomy, physiology, pathology or function, which are present for at least 6 months. Damage may be the result of prior active disease, complications of therapy, co-morbid conditions or other events.

# Introduction - II

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- This work is part of a larger photo-essay of the cutaneous manifestations of the IIM, being prepared by the International Myositis Outcome Assessment Collaborative Study Group (IMOACSG). Please contact the authors for permission to reproduce in any way.

# VAS Scoring of Cutaneous Damage – General Principles

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- The VAS score is an integration of severity of damage based upon all of the cutaneous manifestations present (not all of which may be part of the MDI).
- Maximum value guidelines (10 cm):
  - Calcinosis with extensive subcutaneous exoskeleton resulting in extreme loss of function
- See specific examples for further guidance in scoring. The proposed scoring in the following examples pre-supposes each lesion is the only cutaneous manifestation present. The scoring also takes into account the sequelae of the lesion (such as limitation of motion, scarring, pain, disfigurement).

# Calcinosis

## *Definition:*

Dystrophic calcification observed clinically or by imaging, which involves the skin, subcutaneous tissue, fascia or muscle. Dystrophic calcification occurs at sites of previous injury or damage.

Calcinosis can be clinically or radiographically distinguished into 4 subtypes (*Blane CE et al., 1984, AJR, 142: 397-400*): superficial plaques or nodules (circumscripta), tumoral (universalis), planar, and exoskeleton.

# Calcinosis – Subtypes: Superficial Plaques or Nodules

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## *Definition:*

Relatively small, circumscribed lesions confined to the cutaneous or subcutaneous tissue. Also known as calcinosis circumscripta.

# Calcinosis, Superficial Plaques and Nodules- Example



- Superficial nodules in the posterior thigh of a child with juvenile DM. Note the trans-epidermal elimination of calcium.
- **Proposed MDI VAS Scoring: 3 cm**

# Calcinosis – Subtypes: Tumoral

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## *Definition:*

Large, extensive circumscribed nodules which are often intramuscular, or can be deposited in skin, subcutaneous tissue, or tendons. Can ulcerate if subcutaneous. Also known as calcinosis universalis.

# Calcinosis, Tumoral - Example



Prepared by IMOACSG, 2001

- Deeper, larger, sometimes subcutaneous deposition of calcium. Here multiple lesions involve the trunk, with relatively normal overlying skin, in a boy with juvenile DM.
- Proposed MDI VAS  
Scoring: 4 cm

# Calcinosis – Subtypes: Planar

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## *Definition:*

Linear accumulations of calcinosis that are along the fascial plane of the subcutaneous tissue or muscles.

# Calcinosis, Planar - Example



- **Dystrophic calcification along the fascial plane of the biceps muscle in a child with juvenile DM. This often presents as palpable indurated, well-demarcated linear deposits, most often along the fascial planes of the extremities.**
- **Proposed MDI VAS Scoring: 5 – 6 cm**

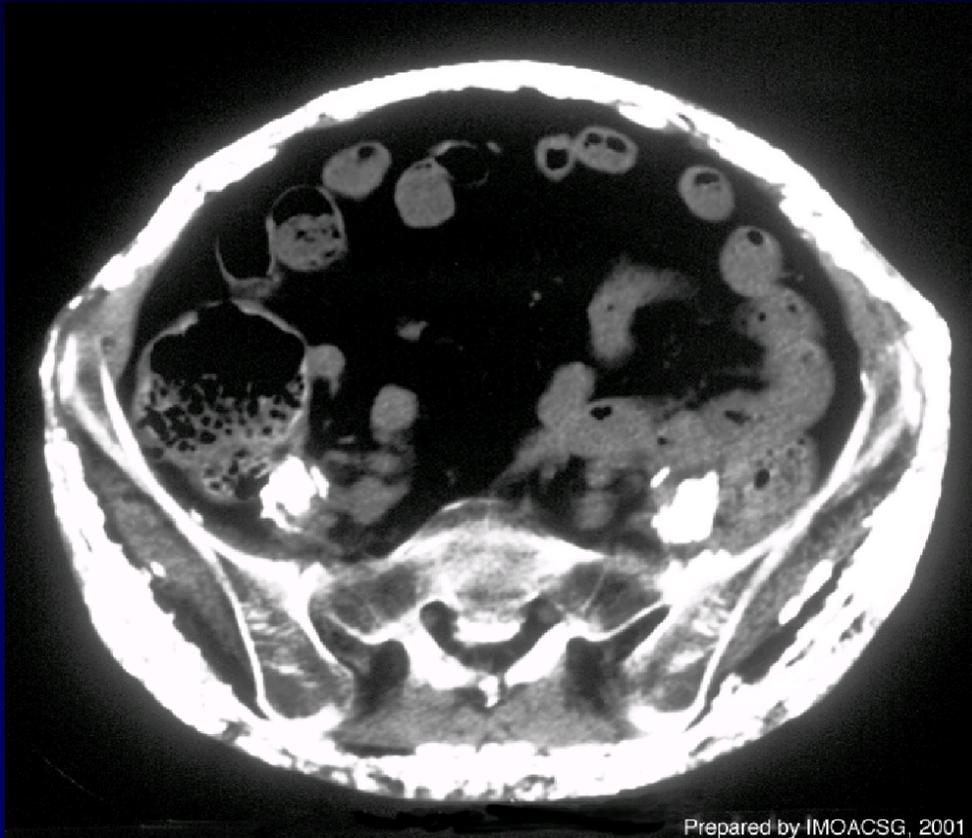
# Calcinosis – Subtypes: Exoskeleton

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## *Definition:*

Widespread sheets of calcium in the muscle, fascia, and subcutaneous tissue, often encasing the patient. Can ulcerate when subcutaneous.

# Calcinosis, Exoskeleton - Example



- Abdominal CT scan demonstrating calcinosis encasing the torso and also involving the psoas muscle in a woman with a history of juvenile DM. The calcium appears as a white density.
- Proposed MDI VAS Scoring: 10 cm

# Alopecia

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## *Definition:*

Hair loss. May be mild or severe, depending on whether it is focal or diffuse, scarring or non-scarring. Scarring alopecia would be considered a more severe manifestation of damage than non-scarring alopecia, because scarring alopecia is more often irreversible.

# Alopecia - Example



- Moderately severe, diffuse alopecia in a young woman with juvenile DM.
- **Proposed MDI VAS Scoring: 5 cm**

# Cutaneous Scarring or Atrophy

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*Definitions:*

Scarring: Permanent damage which results in dermal fibrosis which can be accompanied by atrophy or hypertrophy.

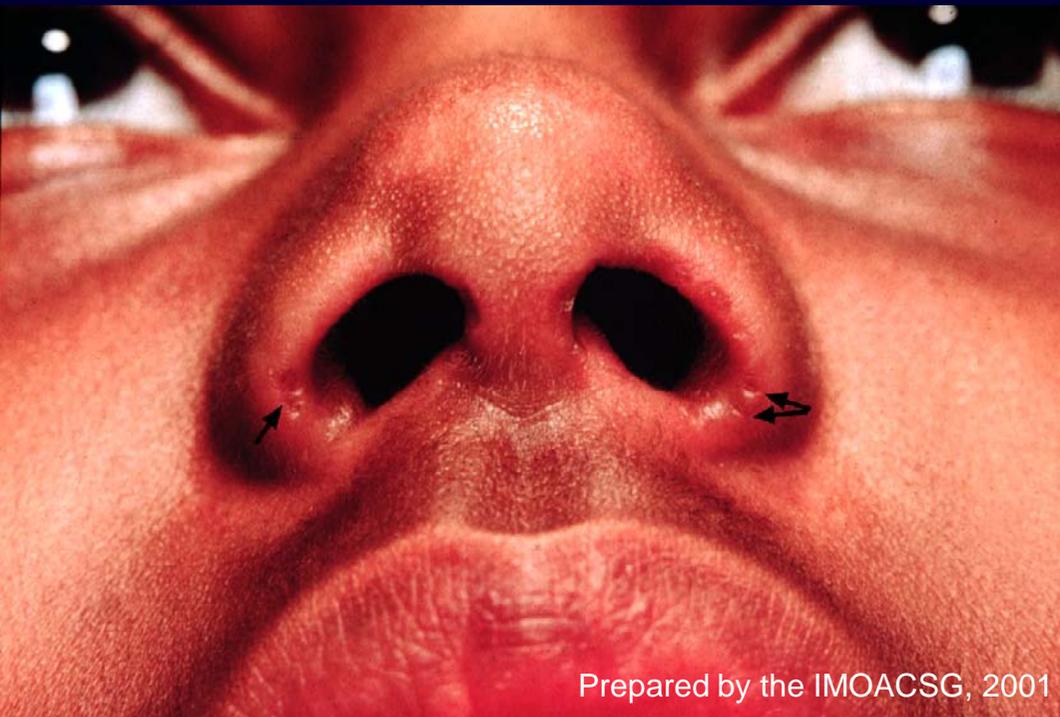
Atrophy: Loss of tissue mass within the epidermis, dermis or subcutaneous fat.

# Cutaneous Atrophy - Example



- Central atrophy presenting as porcelain white depression with telangiectasia involving Gottron's papules of the fingers of a girl with juvenile DM.
- **Proposed MDI VAS Scoring: 3 cm**

# Scarring - Example



Prepared by the IMOACSG, 2001

- Female with juvenile DM who developed shallow encrusted ulcerations along the margins of her nares, which upon healing, left shallow depressed scars with slight loss of tissue bulk.
- **Proposed MDI VAS**  
**Scoring: 1 – 2 cm**

# Poikiloderma

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*Definition:*

Fine speckled pattern of hyper- and hypopigmented macules interspersed with telangiectasia and cutaneous atrophy (requires all 4 features).

# Poikiloderma - Example



- Poikiloderma, mixture of hypo- and hyper-pigmentation with telangiectasia and atrophy, involving both sun-exposed and non sun-exposed areas with sparing of the axillary vault in a boy with juvenile DM.
- **Proposed MDI VAS Scoring: 8 cm**

# Lipodystrophy

## *Definition:*

A disturbance in the distribution of body fat, most frequently manifesting as a loss of subcutaneous fat. This may be **generalized** (widespread) or **partial** (involving only extremities), and can be observed clinically or with imaging studies.

# Lipodystrophy - Example



- Generalized loss of subcutaneous fat along the extremities and trunk in a teenage girl with juvenile DM and generalized lipodystrophy. In the thighs and calves, there is a resulting accentuation of the muscles. She also has acanthosis nigricans in the axilla and neck line from accompanying insulin resistance.
- **Proposed MDI VAS Scoring:**  
8 cm

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