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Firstly described in 2005 in a Japanese cohort patients with **clinically amyopathic dermatomyositis (CADM)**

- Name: Anti-Melanoma Differentiation-Associated Gene 5 antibodies
- **Target antigen:** Recognizes interferon induced with helicase C domain protein 1 (**IFIH1/MD5**)
- Family: **Myositis specific antibodies (MSA)**

Main disease: Dermatomyositis
Prevalence:

- **13 to 35%** of classic DM
- **53% to 73%** of ADM
- **7%** of Juvenile dermatomyositis

Strong association with rapidly-progressive (RP) and severe **interstitial lung disease (ILD)**



High sensitivity (77%) and specificity (86%) to predict RP-ILD in DM patients.
Prognostic markers for RP-ILD.:
-high serum ferritin (SF) levels
-low lymphocyte count
-positive anti- TRIM21(Ro-52) antibodies



Screening technique:

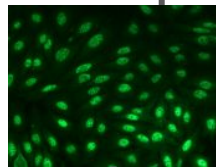
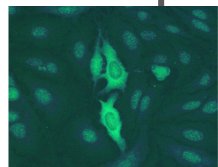
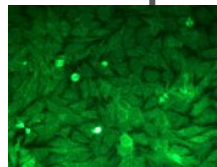
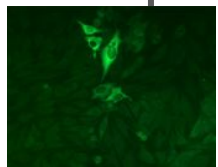
Indirect Immunofluorescence (IIF) on HEp-2 cells:

- Good sensitivity
- Inconstant IIF pattern

**Anti-MDA5
(anti-CADM-140)
antibodies**

3 distinct clinical phenotypes, according to the predominance of pulmonary, skin-articular or vascular symptoms

- **Skin involvement:** skin ulcerations, Gottron's papules, and palmar papules, with severe **vasculopathy** upon biopsy.
- **Muscle involvement:** Mostly **Amyopathic Dermatomyositis** and muscle biopsies are often normal.
- Intermediate risk of cancer according to ACR



Typical fine granular cytoplasmic staining in rare clustered cells

Granular cytoplasmic pattern in all cells

Nuclear speckled pattern, associated with the typical cytoplasmic pattern

Isolated nuclear speckled pattern

Negative IIF patterns do not rule out anti-MDA5 positivity. If the clinical presentation strongly suggests the presence of anti-MDA5 antibodies, additional assays should be performed



Confirmation techniques:

- **Immunoprecipitation (IP)**
- **Line/dot blot immunoassays** (sp: 96-99%, s: 75-93%)
- **ELISA** (sp: 100%, s: 98%)

