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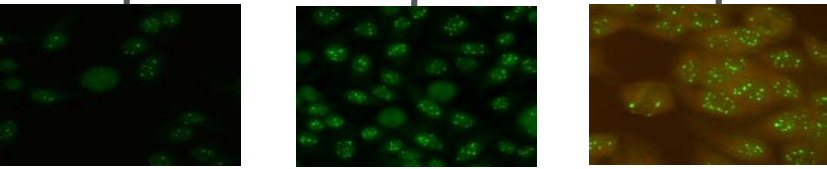
Anti-NXP2 antibodies

- Firstly described in **1997** as an anti-MJ Antibody in a cohort of juvenile dermatomyositis (DM).
- **Antigen target: Nuclear Matrix Protein NXP 2** (also called MORC3): a 140 kDa protein localized in promyelocytic bodies within the nucleoplasm.

- **Family:** Myositis-specific autoantibodies (MSAs)
- Mainly associated with **juvenile DM** with a **high specificity** (95%-98%) and **low sensitivity** (1-1.5%)
- Reported frequency : 1 to 20%** (1-2% in adults)

Screening technique:
Indirect immunofluorescence (IIF) on HEp-2 cells
These aspects are **not always as typical** (may only be detectable at low serum dilutions of less than 1/160) 

IIF pattern on HEp-2 cells: nuclear labeling with a **dot-like appearance** since the target antigen is closely associated with the nuclear matrix




- Confirmation techniques:**
- Line/dot blot immunoassays
 - ELISA
 - laser bead immunoassay (ALBIA)
 - Immunoprecipitation (IP)

Clinical features in dermatomyositis

Severe muscle weakness **Dysphagia** **Interstitial Lung Disease is not common**

Calcinosis ++

- Prognosis:**
- In American College of Rheumatology guidelines of 2022: Considered as a **high-risk factor** in the criterias for **IIM-associated cancer risk**
 - Association with cancer is mostly in adult forms
 - ⇒ **in adult anti-NXP2 IIM: mandatory enhanced cancer screening at diagnosis** 
 - + **Basic screening at 1, 2 and 3 years after IIM onset**
 - No evidence to suggest that the presence of anti-NXP2 Ab confers a poor prognosis with respect to overall survival.

