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- Firstly described in **1980** in patients with CREST syndrome.
- **Family:** Antinuclear antibody "dots" type
- **Target antigen: Centromeres**, (DNA-protein complex) six centromeric nucleoproteins CENP-A ==> CENP-F
CENP-B is the primary autoantigenic protein

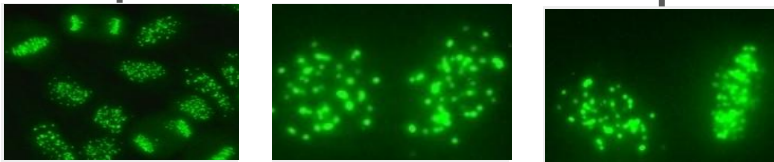
- Main disease: Systemic sclerosis (SSc)**
But also:
- Primary biliary cholangitis (10- 20%)
 - Sjögren's syndrome (3-10%)

Systemic sclerosis (SSc):

- Sensitivity (20%–40%) and the specificity (> 90%)
- Negative predictive value :>90%
- Positive predictive value :20-40%
- Included in the **international 2013 ACR-EULAR classification criteria for SSc** weight: 3 points
- Can be detected years before the onset of the disease and are rarely present in healthy individuals

Screening technique:
Indirect immunofluorescence (IIF) on HEp-2 cells:
Gold standard
The positivity of anti-centromere antibodies can be confirmed by IIF pattern only

IIF pattern on HEp-2 cells: Centromeric, a discrete speckled staining of both interphase nuclei and metaphase chromatin. Discrete coarse speckles (40-80/cell) scattered in interphase cells and aligned at the chromatin mass on mitotic cells.



Anti-centromere antibodies (ACA)

Clinical manifestations:

Subcutaneous calcinosis



Raynaud's syndrome



Telangiectasia

Esophageal involvement



Sclerodactyly

Associated with:

- limited cutaneous forms (80%)
- pulmonary hypertension (20%)
- better prognosis and slower development

Confirmation techniques:

- **Line/dot blot immunoassays:** Specificity 93%-100%.
- **ELISA:** Sensitivity 73-100%
- **Chemiluminescence immunoassay (CIA)**
- **Fluorescence enzyme immunoassay (FEIA)**



