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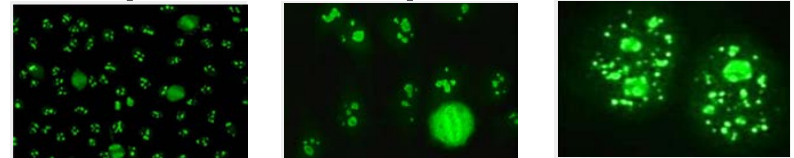
**Anti Fibrillar  
antibodies  
(anti-U3 RNP)**

Firstly described in 1982 in the serum of patients with Systemic Sclerosis (SSc)

**Target antigen: Fibrillarin**, a highly conserved nucleolar protein of 34 kDa, a component of the small nucleolar RNP (snoRNP) complexes containing C/D box RNA sequences snoRNP particles and involved in the processing of ribosomal RNA.  
**Family:** SSc specific antibodies

**Screening technique:** Immunofluorescence (IIF) on HEp-2 cells

**IIF pattern on HEp-2 cells:** Clumpy nucleolar pattern: an irregular, clumpy staining of the nucleoli and with reticular mitosis at the metaphase and telophase plates.



- **Main disease: Systemic sclerosis (SSc)**
  - Prevalence: 4% to 10%
  - Specificity: 95-97%

- Not included in 2013 ACR/ EULAR criteria for SSc
- Found mainly in **male** of **Afro-Caribbean** patients with a **younger** age at diagnosis in comparison with other SSc patients
- Associated with **diffuse cutaneous** SSc more than limited cutaneous SSc
- Frequent **Muscular involvement**
- Higher risk of **gastrointestinal involvement**
- Severe prognosis, due to **pulmonary arterial hypertension**

**Confirmation techniques:**

- **Immunoprecipitation (Gold standard)** complexity of performance and interpretation
- **Western-blot** anti-fibrillar antibodies were difficult to detect
- **Fluorescence enzyme immunoassay (FEIA):**
- **Line/dot blot immunoassays:** based on recombinant fibrillarin, showed a sensitivity of 100% and a specificity of 94.7%. An acceptable and attractive alternative to IP for anti-U3-RNP detection

