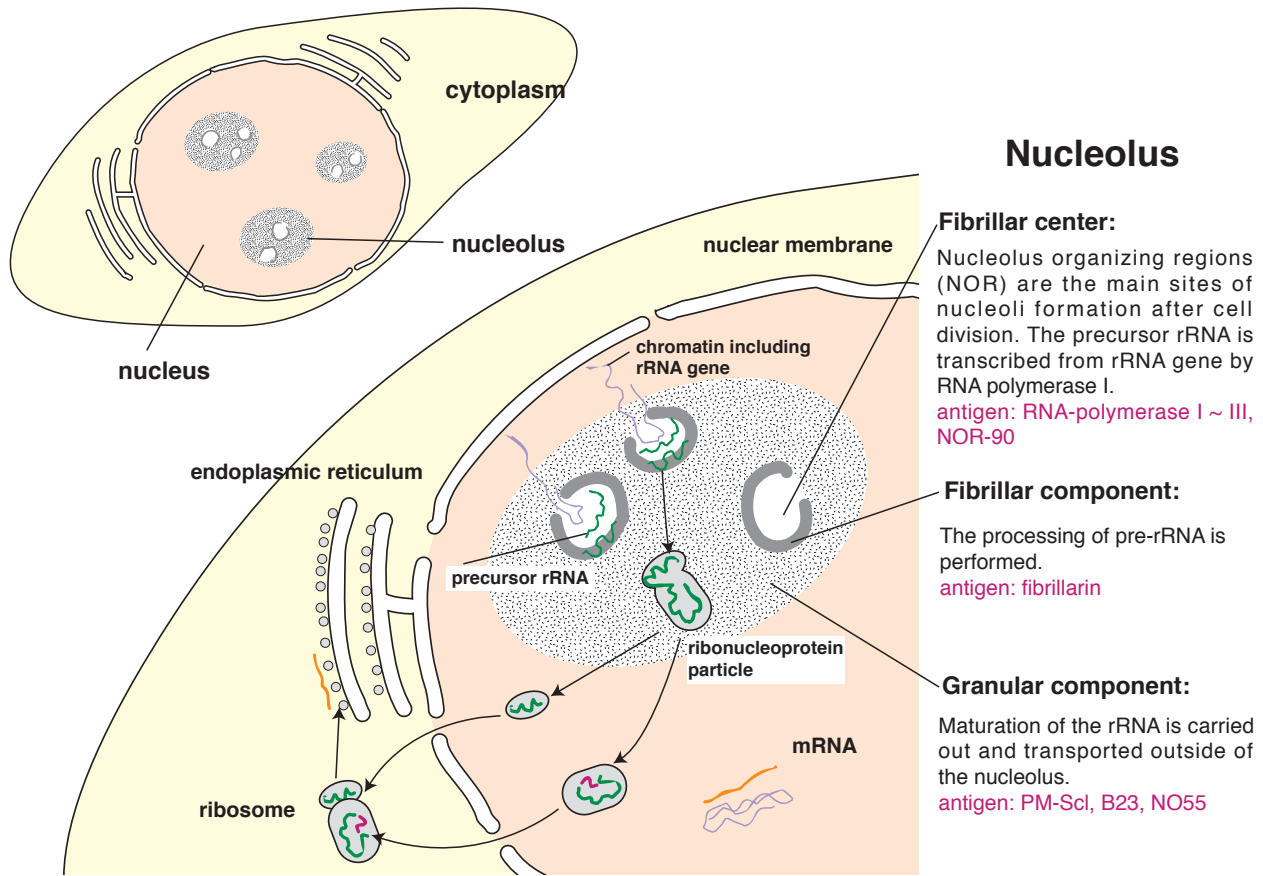


■ Autoantibodies recognizing nucleolus related proteins



The nucleolus is composed of the central fibrillar region and the outer granular component. In the fibrillar center, nucleolus organizer region (NOR) is related to the reformation of the nucleolus after cell division.

Anti-U3RNP antibody (Anti-fibrillarin antibody)

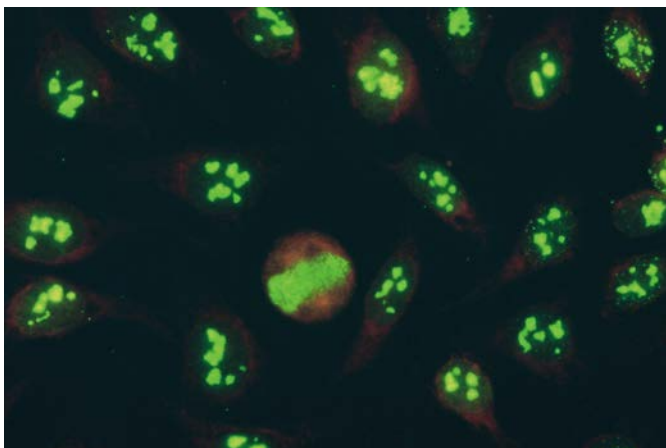


photo 29

Pattern	Clumpy nucleolar staining in interphase cells with chromosomal region fluorescence of mitotic cells.
Antigen	U3RNA-bound protein; fibrillarin (34kD)
Other analytical method	Immunoprecipitation
Clinical significance	These antibodies occur in SSc (mainly observed in patients with diffuse cutaneous scleroderma not associated with arthritis and lung fibrosis), and in hepatocellular carcinoma.
References	29, 30

SLE: systemic lupus erythematosus, SSc: systemic sclerosis, MCTD: mixed connective tissue disease, SS: Sjögren's syndrome, PM/DM: polymyositis/dermatomyositis, RA: rheumatoid arthritis, PBC: primary biliary cirrhosis, AIH: autoimmune hepatitis

Anti-RNA polymerase I antibody

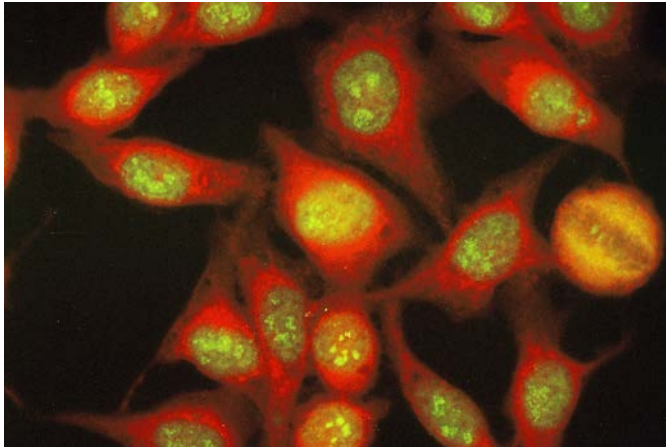


photo 30

Pattern	Punctate or speckled nucleolar staining in interphase cells. The chromosomal regions show no staining in mitotic cells.
Antigen	RNA polymerase I, II and III component proteins (more than 10 proteins ranged from 12.5 to 210kD). The autoantibodies targeting RNA polymerase II or III, and those reacting to more than one RNA polymerase were reported.
Other analytical method	Immunoprecipitation
Clinical significance	These antibodies occur in patients with SSc, frequently associated with patients with severe internal organ involvement (mainly lung and kidney) and poor prognosis.
References	4, 12, 29

Anti-7-2RNP antibody (Anti-Th antibody)

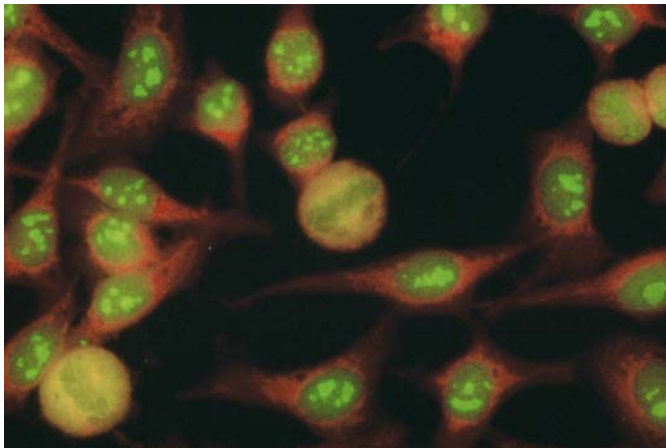


photo 31

Pattern	Dotty nucleolar staining
Antigen	RNaseP(8-2RNP), RNaseMRP(7-2RNP)
Other analytical method	Immunoprecipitation
Clinical significance	These antibodies occur in patients with scleroderma, mainly associated with limited cutaneous involvement.
References	29

Anti-PM-Scl antibody

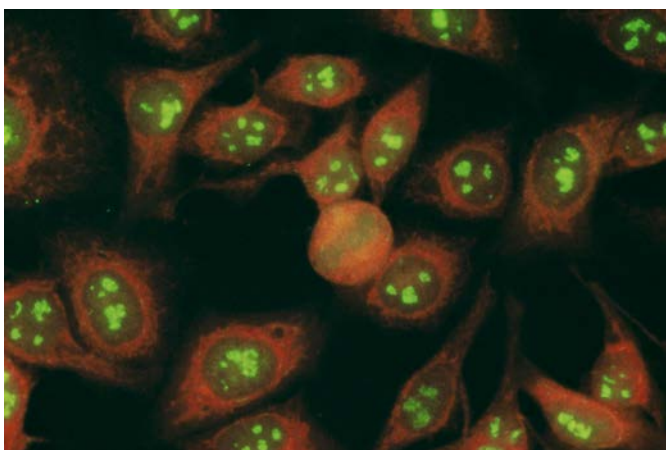


photo 32

Pattern	Homogeneous or diffuse nucleolar staining with no chromosomal staining in mitotic cells.
Antigen	The protein complex of more than 10 nucleolar proteins including 75kD and 100kD polypeptides.
Clinical significance	These antibodies occur in patients with polymyositis/scleroderma overlap syndrome in Europe and America, but not in Japan.
References	29

Anti-NOR-90 antibody

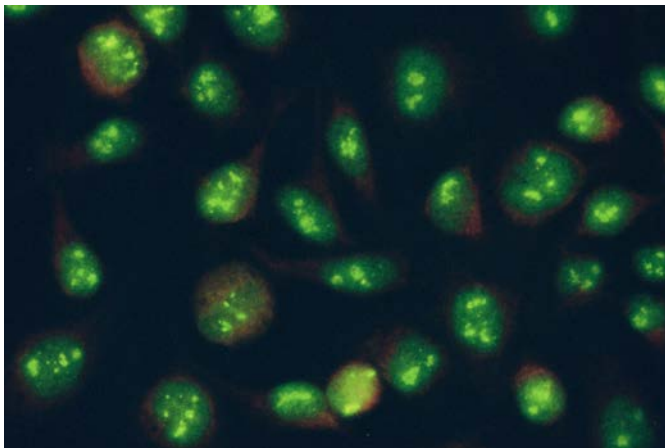


photo 33

Pattern	Coarse speckled or granular nucleolar staining in interphase cells. Several intensely stained dots can be seen in the chromosomal regions in mitotic cells.
Antigen	90kD/92kD proteins in nucleolus organizer region, recognizing RNA polymerase I transcription factor, hUBF (human upstream binding factor) in the fibrillar center.
Other analytical method	DID, ELISA, WB, Immunoprecipitation
Clinical significance	These antibodies occur in patients with SSc frequently associated with Raynaud's phenomenon, also those with hepatocellular carcinoma and SS in Japan.
References	30, 31

Anti-RNA helicase antibody

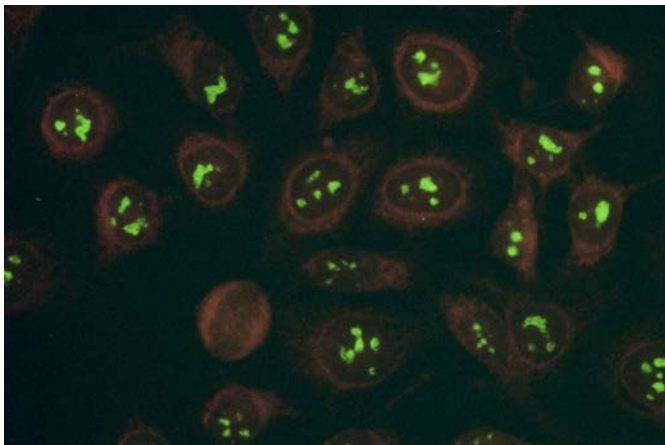


photo 34

Pattern	Dense speckled nucleolar staining with no chromosomal staining in mitotic cells.
Antigen	100 kD nucleolar RNA helicase protein
Other analytical method	Immunoprecipitation
Clinical significance	These antibodies occur in patients with SSc, SLE or watermelon stomach disease.
References	32, 33

Other autoantibodies to nucleolar antigens:

Anti-B23/nucleophosmin antibodies occur in patients with hepatocellular carcinoma and show homogeneous nucleolar staining.

[References: 30, 34]

Anti-No55 antibodies occur in patients with interstitial cystitis and show homogeneous nucleolar pattern. [Reference: 35]

SLE: systemic lupus erythematosus, SSc: systemic sclerosis, MCTD: mixed connective tissue disease, SS: Sjögren's syndrome, PM/DM: polymyositis/dermatomyositis, RA: rheumatoid arthritis, PBC: primary biliary cirrhosis, AIH: autoimmune hepatitis