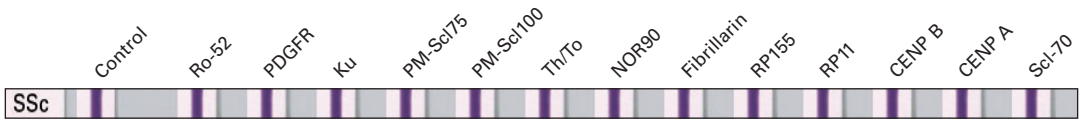
EUROLINE Systemic Sclerosis (Nucleoli) Profile (IgG)



- **Multiplex approach for confirmation and differentiation of nucleolar ANA patterns**
- **Comprehensive spectrum of systemic-sclerosis-associated antigens in one test kit ensures highest possible detection rate**
- **Autoantibodies enable subclassification in limited and diffuse forms of systemic sclerosis and overlap syndrome and also provide information on possible clinical courses**
- **Fully automated incubation and evaluation of immunoblot strips possible via EUROBlotOne**

Technical data

Antigens	Recombinant antigens: CENP A, CENP B, RP11, RP155, fibrillarin, NOR90, Th/To, PM-Scl100 (100 kDa), PM-Scl75 (75 kDa), Ku, PDGFR, Ro-52 (52 kDa) Native antigen: Scl-70
Sample dilution	Serum or plasma, 1 : 101 in sample buffer
Test procedure	30 min / 30 min / 10 min, room temperature, fully automatable
Test kit format	16 membrane strips; kit includes all necessary reagents
Automation	Compatible with all commercial blot processing systems, e.g. EUROBlotOne or EUROBlotMaster from EUROIMMUN
Order number	**DL 1532-1601 G**

Clinical significance

Systemic sclerosis (SSc) belongs to the collagenoses, a group of autoimmune connective tissue diseases. It affects the skin and internal organs. Around 2 to 50 out of 100,000 people suffer from SSC worldwide (USA: 25 out of 100,000). The incidence amounts to 12 new cases per 100,000 people per year. The disease occurs mainly in middle adulthood. Women are affected three to four times more frequently than men. Black people have a greater risk of acquiring the disease. A higher frequency among members of one family is rare. Early symptoms of SSc are shortening of the lingual frenum and Raynaud's syndrome (1. stage: ischemia of the hands and feet with numbness and pains, 2. stage: local cyanosis caused by hypoxia, 3. stage: reactive hyperaemia with redness, prickling and throbbing). In the following phase oedema of the hands and feet develop. The skin becomes stiff and in later stages atrophic, waxy and thin. Finally, deformation of the hands occurs. The fingers become fixed in a bent position (claw hand) and are highly tapered at the ends (Madonna fingers). Furthermore, the characteristic mask-like face with rigid mimic develops, leading to microstomia (reduced capability of opening the mouth) and problems in closing the eye lids. Finally, callosity of the inner organs, particularly of the digestive tract, lungs, heart and kidneys occurs. SSC is divided into limited and diffuse forms, depending on the cutaneous distribution. In the limited form, skin involvement is limited to the distal extremities. In the diffuse form (also proximal systemic sclerosis) the symptoms are diffusely distributed over the trunk, the proximal and distal extremities and the face. The so-called CREST syndrome with calcinosis, Raynaud's syndrome, oesophageal dysfunction, sclerodactyly (thin, pale, thickened and hairless skin on the fingers) and teleangiectasias (persisting pathological dilation of superficial skin vessels) is a special subform of SSc. The connective tissue of the lungs, kidneys, oesophagus and heart is particularly at risk. At present, lung involvement is the most frequent cause of death from SSc. Manifest SSc is the collagenosis with the highest vital risk for the patient. Since SSc presents under various forms and on different body parts and may even come to a standstill, clinical diagnosis is difficult.

Diagnostic application

The newly developed EUROLINE Systemic Sclerosis (Nucleoli) Profile (IgG) is the first membrane-based test system to provide such a wide range of systemic sclerosis-associated antigens. Thus the test allows differentiation of antibodies with nucleolar patterns obtained in indirect immunofluorescence. The assay surpasses all existing test methods with respect to sensitivity and specificity and provides an effective and accurate determination of autoantibodies in systemic sclerosis and overlapping syndrome. New studies have shown that subclassification and substratification of the disease by means of autoantibodies is useful, especially in the early stages. It supplements the wide range of EUROLINE products, which can be performed fully automatically using user-friendly automation solutions and evaluation software.

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Test principle

The EUROLINE is a qualitative in vitro immunoassay, in which membrane strips printed with lines of purified, biochemically characterised antigens are used as solid phase. Each antigen is coated onto a separate membrane fragment, enabling the production process and thereby the efficiency of antibody detection to be optimised for each protein. Since antigen bands are located at defined positions, results can be evaluated visually without the need for additional equipment. Correct performance of all test steps is confirmed by staining of the control band.

Automatic processing

EUROBlotOne is a fully automatic device for the standardised processing of EUROIMMUN line assays (EUROLINE, EUROLINE-WB, Westernblot) – from sample recognition to the final test result. Samples are pipetted by the device and all incubation and washing steps are carried out automatically. Finally the data of the pictures taken by the integrated camera are automatically evaluated and digitally archived by the EUROLineScan software. Alternatively, the immunoblot strips can be incubated by the EUROBlotMaster and scanned using the EUROBlotScanner or photographed directly in the incubation tray using the EUROBlotCamera. Also in this case, the automatic evaluation is carried out by the EUROLineScan software. The bidirectional communication with a laboratory information management system for import of work lists and export of results is enabled by EUROLineScan or, optionally, the laboratory management software EUROLabOffice. A separate results sheet can be produced for each patient.

Correlation

129 sera from patients with clinically characterised SSc (limited and diffuse form) and 142 sera from control patients (50 polymyositis/dermatomyositis (PM/DM), 50 systemic lupus erythematosus (SLE), 42 rheumatoid arthritis (RA)) were tested using the indirect immunofluorescence test (IIFT) based on HEp-2 cells (EUROIMMUN). 92.8% (90/97) of the SSc sera with a nucleolar pattern reacted positively with the EUROLINE Systemic Sclerosis (Nucleoli) Profile (IgG). But only 17.6% (3/17) of the nucleolar pattern sera from the control panels showed a positive reaction. This shows that identification and differentiation of a high number of SSc-specific and -associated antibodies is possible.

Antibodies against	Indirect immunofluorescence pattern SSc patients (n = 129)					Control panels (n = 142)
	Nucleolar (n = 97)	Centromere (n = 12)	Nucleo./ Centrom. (n = 2)	Other (n = 17)	Negative (n = 1)	Nucleolar (n = 17)
Scl-70	80	–	1	2	–	–
CENP A	1	12	1	–	–	–
CENP B	1	12	1	–	–	–
RP11	6	–	–	–	–	–
RP155	7	1	–	–	–	–
Fibrillarin	1	–	–	–	–	–
NOR90	4	–	–	–	–	–
Th/To	8	–	–	–	–	1
PM-Scl100	5	–	–	3	–	2
PM-Scl75	12	1	–	4	–	1
Ku	4	–	–	–	–	–
PDGFR	0	–	1	–	–	–
Positive reaction with at least one SSc antigen	**90**	**12**	**1**	**7**	**0**	**3***

*Sera from dermatomyositis/polymyositis panel

Sensitivity and specificity

129 sera from patients with clinically characterised SSc (limited and diffuse form) and 142 sera from control patients (50 PM/DM, 50 SLE, 42 RA) and 60 sera from healthy blood donors were investigated using the EUROLINE Systemic Sclerosis (Nucleoli) Profile (IgG). The total detection rate in the systemic sclerosis panel was 85.3%.

	EUROIMMUN EUROLINE Systemic Sclerosis (Nucleoli) Profile (IgG)											
Autoantibodies against	**Scl-70**	**CENP A**	**CENP B**	**RP11**	**RP155**	**Fibrillarin**	**NOR90**	**Th/To**	**PM-Scl100**	**PM-Scl75**	**Ku**	**PDGFR**
Sensitivity (n = 129)	65%	11%	13%	5%	7%	2%	4%	6%	7%	12%	6%	1%
Specificity (n = 202)	99%	99%	99%	100%	100%	100%	99%	98%	99%	98%	99%	100%

Autoantibodies against Ro-52 are not specific for SSc, but they frequently occur together with autoantibodies against SSc-specific antigens.

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