



Product Datasheet

Product Name	PM/Scl 100 Human Recombinant
Cata No	CB501064
Source	<i>Sf9 insect cells.</i>
Synonyms	Exosome component 10, Polymyositis/scleroderma autoantigen 2, Autoantigen PM/Scl 2, Polymyositis/scleroderma autoantigen 100 kDa, PM/Scl-100, P100 polymyositis-scleroderma overlap syndrome-associated autoantigen, EXOSC10, PMSCL, PMSCL2, p2, p3, p4, RRP6, Rrp6p, PM-Scl.

Description

PM/Scl 100 is a nucleolar protein and represents one crucial component of the so-called "PM/Scl complex" or "nuclear exosome", which is an RNA-processing complex consisting of a number of exoribonucleases (not to be confused with certain multivesicular bodies which are shed by cultured cells into the culture fluid and are also called exosomes).

Autoantibodies against PM/Scl 100 occur in patients suffering from polymyositis, scleroderma or polymyositis/scleroderma overlap syndromes. While antibodies to other components of the exosome complex (PM/Scl 75, several Rrp proteins) can be found in some sera, practically all patients with autoantibodies against the PM/Scl complex are positive for PM/Scl 100 antibodies, making this specificity an important marker. The presence of PM/Scl 100 autoantibodies is a comparatively "good" prognostic sign in polymyositis/scleroderma overlap syndrome unlike the poor prognosis seen when other myositis and systemic sclerosis specific

autoantibodies are present.

PM/Scl 100 Human Recombinant produced in SF9 is a glycosylated, polypeptide chain having a molecular mass of 120 kDa.

PM/Scl 100 is expressed with a -6xHis tag and purified by proprietary chromatographic techniques.

Physical Appearance

Sterile Filtered clear solution.

Purity

Greater than 70% as determined by SDS-PAGE.

Formulation

PM/Scl 100 is supplied at a concentration of 0.8mg/ml in 20mM HEPES, pH-7.5, 100mM NaCl, 0.01mM EDTA, 0.02% SDS.

Applications

Western-Blot with monoclonal anti-hexa-His-tag antibody & polymyositis patient sera.