

Desmin (ABT19R) rabbit mAb

Catalog No :	YM7110
Reactivity :	Human;Mouse;(predicted: Rat)
Applications :	IHC;WB; ELISA
Target :	Desmin
Fields :	>>Hypertrophic cardiomyopathy;>>Arrhythmogenic right ventricular cardiomyopathy;>>Dilated cardiomyopathy
Gene Name :	DES
Protein Name :	Desmin
Human Gene Id :	1674
Human Swiss Prot No :	P17661
Immunogen :	Synthesized peptide derived from human Desmin AA range:400-470
Specificity :	This antibody detects endogenous levels of Desmin
Formulation :	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source :	Monoclonal, Rabbit IgG1, Kappa
Dilution :	IHC 1:100-500, WB 1:500-1000, ELISA 1:5000-20000
Purification :	Recombinant Expression and Affinity purified
Storage Stability :	-15°C to -25°C/1 year(Do not lower than -25°C)
Molecularweight :	54kD
Background :	This gene encodes a muscle-specific class III intermediate filament. Homopolymers of this protein form a stable intracytoplasmic filamentous network connecting myofibrils to each other and to the plasma membrane. Mutations in

this gene are associated with desmin-related myopathy, a familial cardiac and skeletal myopathy (CSM), and with distal myopathies. [provided by RefSeq, Jul 2008],

Function :

disease:Defects in DES are the cause of cardiomyopathy dilated type 1I (CMD1I) [MIM:604765]. Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.,disease:Defects in DES are the cause of desmin-related cardio-skeletal myopathy (CSM) [MIM:601419]; also known as desmin-related myopathy (DRM). CSM is characterized by skeletal muscle weakness associated with cardiac conduction blocks, arrhythmias, restrictive heart failure, and by intracytoplasmic accumulation of desmin-reactive deposits in cardiac and skeletal muscle cells. A desmin-related myopathy can have a distal onset, it is then known as hereditary distal myopathy (HDM).,disease:Defects in DES are the cause of neurogenic scapuloperoneal syndrome Kaeser type (Kaeser syndrome) [MIM:181400].

Subcellular Location :

Cytoplasmic

Expression :

Cytoplasmic

Products Images