

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:

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

P17661

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

Tag: hot,recombinant

Sort: 25005

No4:

Host: Rabbit

Modifications: Unmodified

Products Images

2/2