

Mouse Anti-Desmin Monoclonal Antibody

Cat. Number:	BH0195
Target Protein:	Human Desmin
Quantity Size:	50ul / 100ul
Clonality:	Monoclonal
Clone No.:	4B12
Isotype:	IgG1
Purity:	≥95%, Purified by Protein G
Form:	Liquid
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Application:	IHC (1:100-500)

Background: Desmin is a muscle-specific, type III intermediate filament that integrates the sarcolemma, Z disk, and nuclear membrane in sarcomeres and regulates sarcomere architecture. In adult striated muscle they form a fibrous network connecting myofibrils to each other and to the plasma membrane from the periphery of the Z line structures. Defects in Desmin are the cause of desmin related cardio skeletal myopathy (CSM) 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). Defects in Desmin are also the cause of dilated cardiomyopathy type 1I (CMD1I). CMD1I is an autosomal form of dilated cardiomyopathy characterized by ventricular dilatation and impaired systolic function. Antidesmin antibodies are useful in identification of tumours of myogenic origin.