

## BACKGROUND

Desmin, a protein of 52 kDa has been identified as the constitutive subunit of class-III intermediate filaments in all muscle tissues, cardiac, skeletal, and smooth. 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 it is encoded by a single copy gene. It is one of earliest known myogenic markers both in heart and in somites. During development Desmin expression precedes all the other muscle-specific structural genes and the myogenic HLH transcription factors myoD, Myogenin, and MRF4, with the exception of Myf-5, suggesting that it might play some modulating role in myogenic commitment and differentiation.<sup>1</sup> In addition, Desmin might be involved in signal transduction and transport processes between the cell surface and the nucleus, which is supported by several pieces of recent information, including its position in the cell where it interlinks the sarcolemma with the nuclear envelope by specifically binding to Ankyrin, other components of the Costameres, and Lamin B, respectively. Lamin B binds to the nuclear matrix-associated regions (MARS) which can activate gene transcription by binding to the nuclear matrix.<sup>2</sup> Furthermore, it was reported that Lamin A and Desmin can also specifically bind MARS in vitro. On the other hand, intermediate filaments serve both as sites of attachment as well as major substrates of several kinases that seem to be involved in signal transduction. In addition, Desmin may be important in mitochondria function. When Desmin is not functioning properly there is improper mitochondrial distribution, number, morphology and function.<sup>3</sup> Mutations in this gene are associated with Desmin-related myopathy, a familial cardiac and skeletal myopathy (CSM), and with distal myopathies.<sup>4</sup>

### References:

1. Weitzer, G. et al: Dev. Biol. 172:422-439, 1995
2. Costa, M.L. et al: Braz. J. Med. Biol. Res. 37:1819-30, 2004
3. Bar, H. et al: J. Struct. Biol. 148:137-52, 2004
4. Goldfarb, L.G. et al: Brain 127:723-34, 2004

## TECHNICAL INFORMATION

### Source:

Desmin Antibody is a mouse monoclonal antibody raised against purified recombinant human desmin fragments expressed in *E. coli*.

### Specificity and Sensitivity:

This antibody detects endogenous desmin proteins without cross-reactivity with other related proteins.

**Storage Buffer:** PBS and 30% glycerol

### Storage:

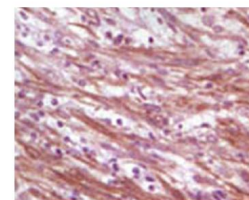
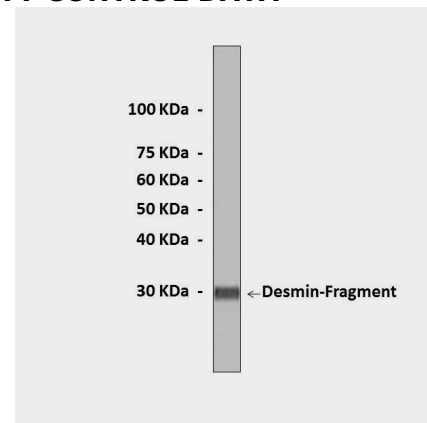
Store at -20°C for at least one year. Store at 4°C for frequent use. Avoid repeated freeze-thaw cycles.

## APPLICATIONS

Application:	*Dilution:
WB	1:1000
IP	n/d
IHC	1:200
ICC	n/d
FACS	n/d

*\*Optimal dilutions must be determined by end user.*

## QUALITY CONTROL DATA



**Top:** Western Blot detection of Desmin proteins in bacterial lysate containing GST-human Desmin fragment fusion proteins (predicted MW 29 kDa) using Desmin Antibody. **Bottom:** This antibody stains paraffin-embedded human muscle sarcoma tissue in immunohistochemical analysis.

