



Mouse anti-Mouse Telethonin monoclonal antibody (CABT-B9335)

This product is for research use only and is not intended for diagnostic use.

PRODUCT INFORMATION

Immunogen	Mouse Telethonin aa. 1-167
Isotype	IgG1
Source/Host	Mouse
Species Reactivity	Mouse, Human, Rat
Purification	The monoclonal antibody was purified from tissue culture supernatant or ascites by affinity chromatography.
Conjugate	Unconjugated
Applications	WB
Format	Liquid
Concentration	250 µg/ml
Size	50 µg
Buffer	Aqueous buffered solution containing BSA, glycerol, and ≤0.09% sodium azide.
Storage	Store undiluted at -20°C.

BACKGROUND

Introduction	Autosomal recessive limb-girdle muscular dystrophy (AR LGMD), a genetically heterogeneous group of disorders affecting the proximal musculature, has eight distinct forms: 2A to 2H. Each of these forms is caused by specific gene mutations. A mutation in the telethonin gene causes
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LGMD 2G. Telethonin, a 19 kDa sarcomeric protein found in striated and cardiac muscle, has a developmentally and functionally regulated expression pattern. Telethonin transcript down-regulation occurs in response to muscle denervation. Protein levels of telethonin are partially regulated by neuronal activity, thereby linking telethonin to dynamic control of myofibrillogenesis and muscle turnover in human skeletal muscle. The telethonin transcript exhibits a pattern of accumulation typical of contractile proteins, suggesting a role for the protein in myofibrillar assembly. Immunofluorescence images show that telethonin co-localizes with myosin, which is an expression pattern typical of sarcomeric proteins. Telethonin has a known sarcomeric binding partner, titin, and is also known as titin CAP (TCAP). Therefore, telethonin plays a major role in AR LGMD 2G and is also important as a developmentally and functionally regulated sarcomeric protein.

Keywords

TCAP; titin-cap; TELE; CMD1N; T-cap; LGMD2G; telethonin; telethonin; titin cap protein; 19 kDa sarcomeric protein; limb girdle muscular dystrophy 2G (autosomal recessive);

GENE INFORMATION

Entrez Gene ID

[8557](#)

UniProt ID

[A2TDC0](#)
