内核膜蛋白 MAN1 抗体

产品货号： mlR18643

英文名称： MAN1

中文名称： 内核膜蛋白 MAN1 抗体

别　　名： Inner nuclear membrane protein Man1; LEM domain containing protein 3; LEM domain-containing protein 3; LEMD3; MAN1_HUMAN.

研究领域： 肿瘤　细胞生物　发育生物学　信号转导　转录调节因子　表观遗传学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Rabbit,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 100kDa

细胞定位： 细胞核

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human MAN1:401-500、911

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed： PubMed

产品介绍： This locus encodes a LEM domain-containing protein. The encoded protein functions to antagonize transforming growth factor-beta signaling at the inner nuclear membrane. Two transcript variants encoding different isoforms have been found for this gene. Mutations in this gene have been associated with osteopoikilosis, Buschke-Ollendorff syndrome and melorheostosis.[provided by RefSeq, Nov 2009]

Function:

Can function as a specific repressor of TGF-beta, activin, and BMP signaling through its interaction with the R-SMAD proteins. Antagonizes TGF-beta-induced cell proliferation arrest.

Subcellular Location:

Nucleus inner membrane.

Tissue Specificity:

Heart, brain, placenta, lung, liver and skeletal muscle.

DISEASE:

Defects in LEMD3 are the cause of Buschke-Ollendorff syndrome (BOS) [MIM:166700]; also known as dermatoosteopoikilosis or disseminated dermatofibrosis with osteopoikilosis or dermatofibrosis lenticularis disseminata with osteopoikilosis or osteopathia condensans disseminata. BOS refers to the association of osteopoikilosis with disseminated connective-tissue nevi. Osteopoikilosis is a skeletal dysplasia characterized by a symmetric but unequal distribution of multiple hyperostotic areas in different parts of the skeleton. Both elastic-type nevi (juvenile elastoma) and collagen-type nevi (dermatofibrosis lenticularis disseminata) have been described in BOS. Skin or bony lesions can be absent in some family members, whereas other relatives may have both.

Defects in LEMD3 are a cause of melorheostosis (MEL) [MIM:155950].

Melorheostosis is a rare mesenchymal dysplasia and one of the sclerosing bone disorders. It is caused by a developmental error, with a sclerotomal distribution, frequently involving one limb. It may be asymptomatic, but pain, stiffness with limitation of motion, leg-length discrepancy and limb deformity may occur.

Similarity:

Contains 1 LEM domain.

SWISS:

Q9Y2U8

Gene ID:

23592

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.