SunLong Biotech Co.,LTD
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Rabbit Anti-Desmin antibody

SL1026R

Product Name	Desmin
Chinese Name	结蛋白抗体
Alias	DESM_HUMAN; DES;
Research Area	Tumour Cardiovascular immunology Signal transduction Cell type markers
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Human Mouse Rat
Applications	WB=1:500-5000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:50-200,IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	52kDa
Cellular localization	cytoplasmic
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human Desmin: 261-360/470
Lsotype	IgG
Purification	affinity purified by Protein A
Buffer Solution	Human,Mouse,Rat1M TBS(pH7.4) with 1% BSA, Human,Mouse,Rat3% Proclin300 and 50% Glycerol.
Storage	Shipped at 4℃. Store at -20 ℃ for one year. Avoid repeated freeze/thaw cycles.
Attention	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
PubMed	PubMed
Product Detail	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.

Function:
Desmin are class-III intermediate filaments found in muscle cells. 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.

Subunit:
Homopolymer. Interacts with DST. Interacts with MTM1.

Subcellular Location:
Cytoplasm.

Post-translational modifications:
ADP-ribosylation prevents ability to form intermediate filaments.

DISEASE:
Defects in DES are the cause of myopathy myofibrillar type 1 (MFM1) [MIM:601419]. A neuromuscular disorder characterized by skeletal muscle weakness associated with cardiac conduction blocks, arrhythmias, restrictive heart failure, and by myofibrillar destruction with intracytoplasmic accumulation of desmin-reactive deposits in cardiac and skeletal muscle cells. Note=Mutations in the DES gene are associated with a variable clinical phenotype which encompasses isolated myopathies, pure cardiac phenotypes (including dilated cardiomyopathy, restrictive cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy), cardiac conduction disease, and combinations of these disorders. If both cardiologic and neurologic features occur, they can manifest in any order, as cardiologic features can precede, occur simultaneously with, or follow manifestation of generalized neuromuscular disease (PubMed:19879535).
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.
Defects in DES are the cause of neurogenic scapuloperoneal syndrome Kaeser type (Kaeser syndrome) [MIM:181400]. Kaeser syndrome is an autosomal dominant disorder with a

peculiar scapuloperoneal distribution of weakness and atrophy. A large clinical variability is observed ranging from scapuloperoneal, limb grindle and distal phenotypes with variable cardiac or respiratory involvement. Facial weakness, dysphagia and gynaecomastia are frequent additional symptoms. Affected men seemingly bear a higher risk of sudden, cardiac death as compared to affected women. Histological and immunohistochemical examination of muscle biopsy specimens reveal a wide spectrum of findings ranging from near normal or unspecific pathology to typical, myofibrillar changes with accumulation of desmin.

Similarity:
Belongs to the intermediate filament family.

SWISS:
P17661

Gene ID:
1674

Database links:

Entrez Gene: 1674 Human

Entrez Gene: 13346 Mouse

Entrez Gene: 64362 Rat

Omim: 125660 Human

SwissProt: P17661 Human

SwissProt: P31001 Mouse

SwissProt: P48675 Rat

Unigene: 594952 Human

Unigene: 6712 Mouse

Unigene: 39196 Rat

Desmin 在很多哺乳动物中的横纹肌和各种平滑肌及其来源的 Tumour 组织中都有表达。结蛋白是一种中间丝蛋白，广泛分布于骨骼肌细胞、平滑肌细胞、心肌细胞和肌 epithelial cells 及其 Tumour 中，主要用于子宫、皮肤、胃肠道及其它横纹肌肉瘤

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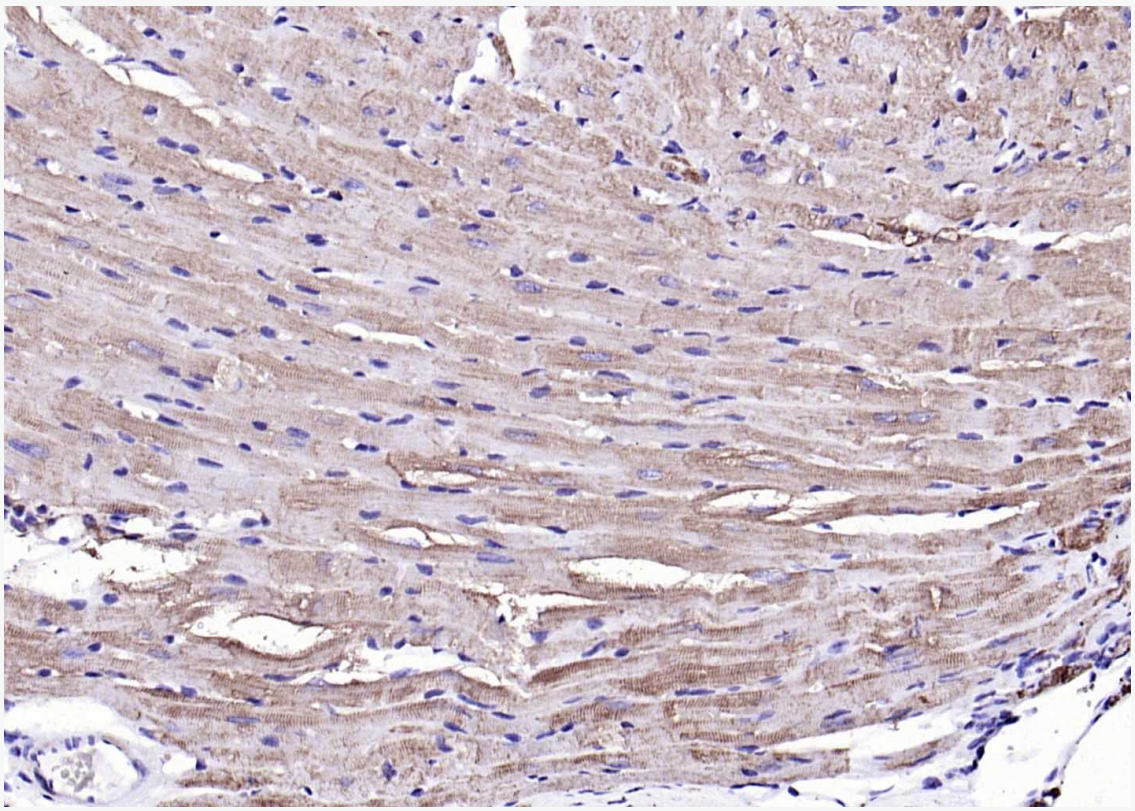
和肌上皮瘤的诊断和鉴别诊断。

Product Picture



Paraformaldehyde-fixed, paraffin embedded (mouse heart); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Desmin) Polyclonal Antibody, Unconjugated (SL1026R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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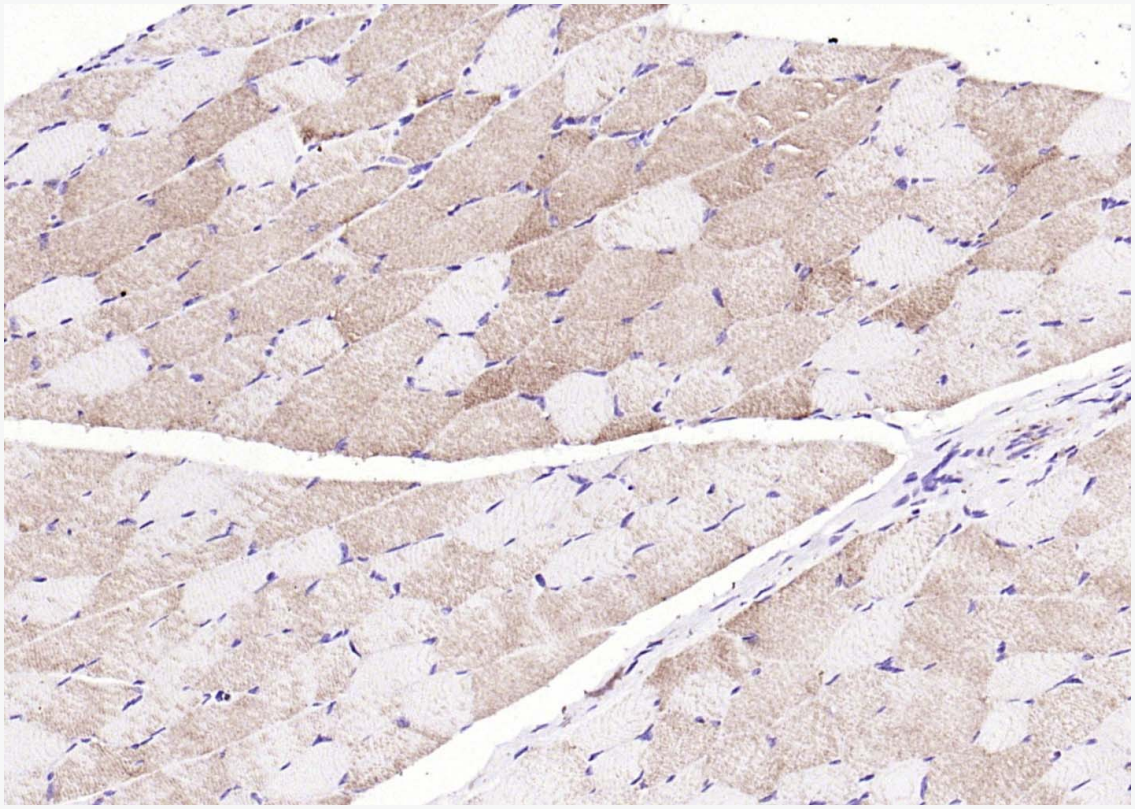

Paraformaldehyde-fixed, paraffin embedded (rat skeletal muscle); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Desmin) Polyclonal Antibody, Unconjugated (SL1026R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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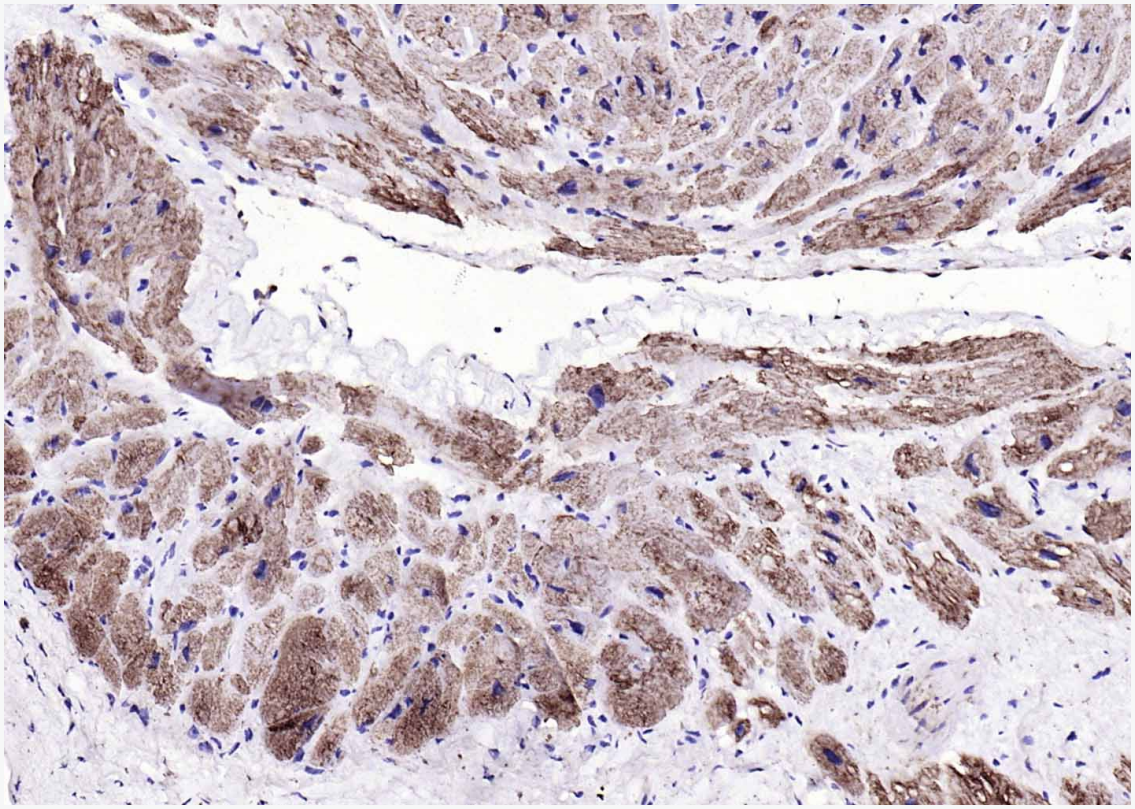

Paraformaldehyde-fixed, paraffin embedded (human myocardium); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Desmin) Polyclonal Antibody, Unconjugated (SL1026R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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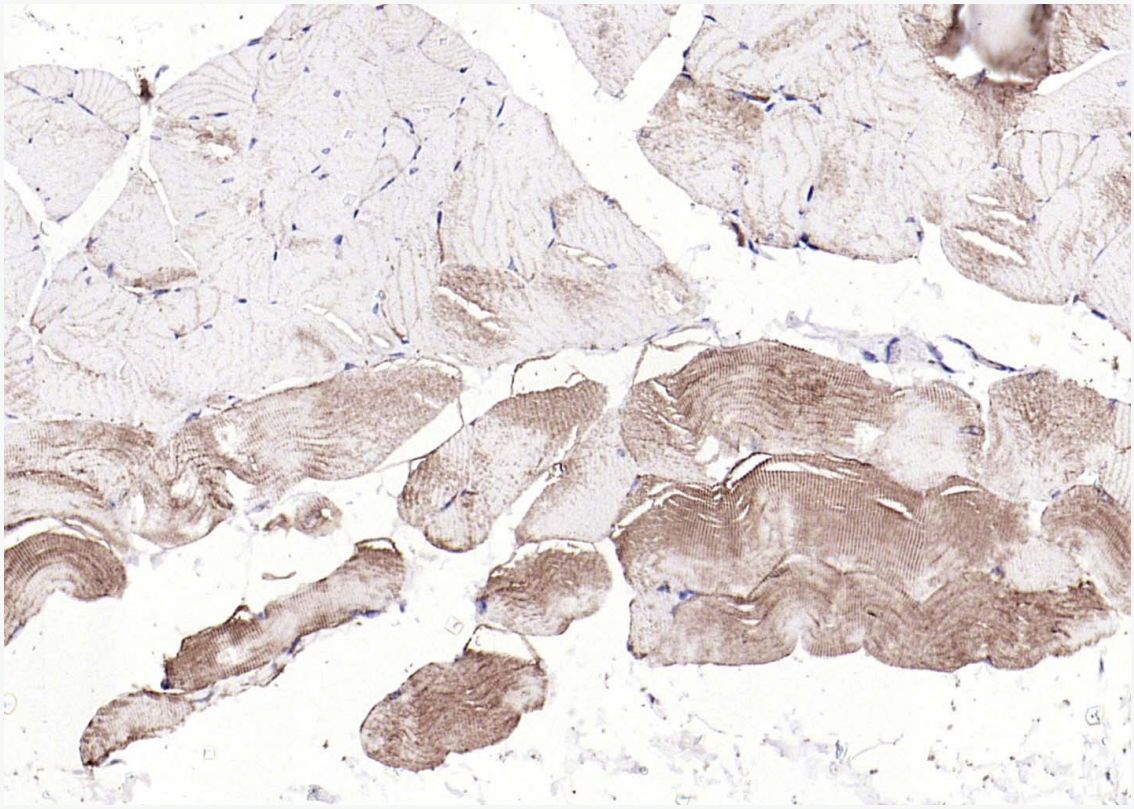

Paraformaldehyde-fixed, paraffin embedded (mouse skeletal muscle); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Desmin) Polyclonal Antibody, Unconjugated (SL1026R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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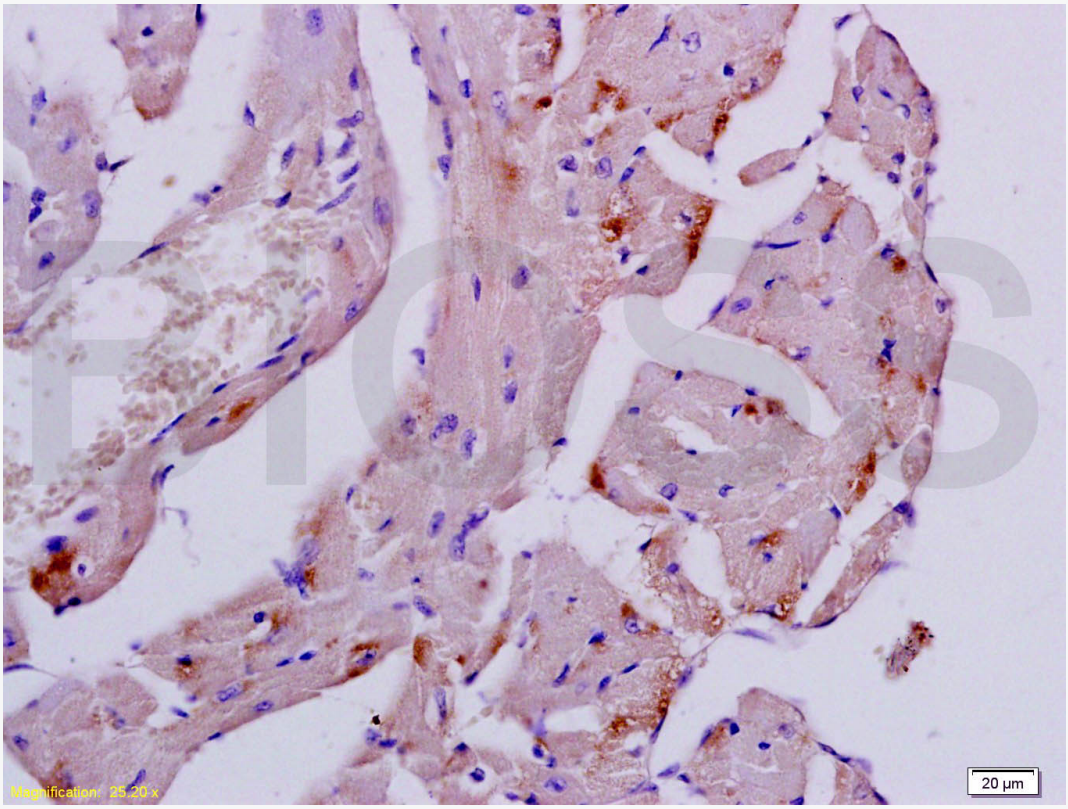

Tissue/cell: mouse heart tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (Human,Mouse,Rat1M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-Desmin Polyclonal Antibody, Unconjugated(SL1026R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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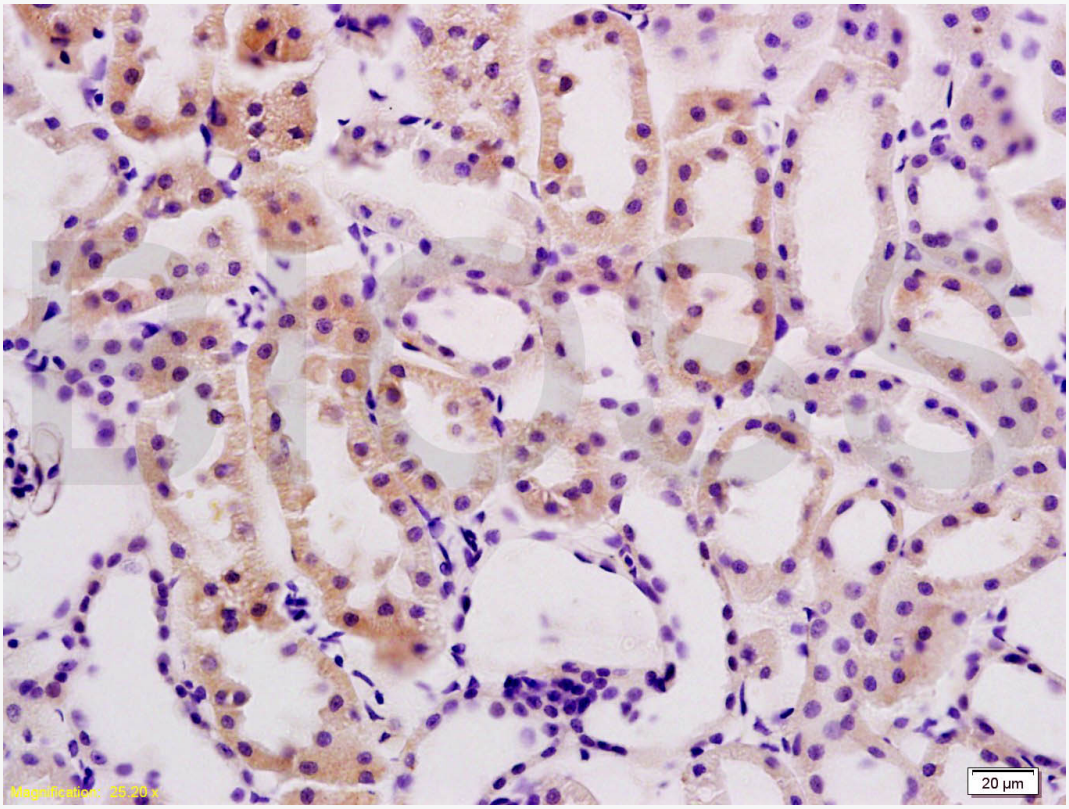

Tissue/cell: rat kidney tissue; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (Human,Mouse,Rat1M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-Desmin Polyclonal Antibody, Unconjugated(SL1026R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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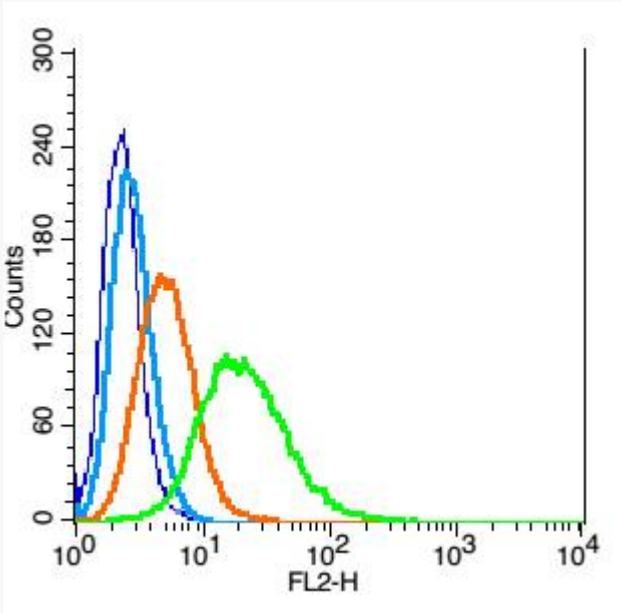

Blank control: Hela(blue).

Primary Antibody:Rabbit Anti- Desmin antibody(SL1026R), Dilution: 1μg in 100 μL 1X PBS containing 0.5% BSA;

Isotype Control Antibody: Rabbit IgG(orange) ,used under the same conditions);

Secondary Antibody: Goat anti-rabbit IgG-PE(white blue), Dilution: 1:200 in 1 X PBS containing 0.5% BSA.

Protocol

The cells were fixed with 2% paraformaldehyde (10 min) , then permeabilized with 90% ice-cold methanol for 30 min on ice. Antibody (SL1026R, 1μg /1x10^6 cells) were incubated for 30 min on the ice, followed by 1 X PBS containing 0.5% BSA + 1 0% goat serum (15 min) to block non-specific protein-protein interactions. Then the Goat Anti-rabbit IgG/PE antibody was added into the blocking buffer mentioned above to react with the primary antibody of SL1026R at 1/200 dilution for 30 min on ice.

Acquisition of 20,000 events was performed.