

Product name:	Sarcoglycan α Rabbit Polyclonal Antibody
Cat number:	ABN17606
Conjugate:	Unconjugated
Size:	100 μ L
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human SGCA. AA range:161-210
Reactivity:	Human,Mouse
Applications:	WB 1:500-1:2000,IHC 1:50-1:300
Molecular Weight:	43kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

sarcoglycan alpha(SGCA) Homo sapiens This gene encodes a component of the dystrophin-glycoprotein complex (DGC), which is critical to the stability of muscle fiber membranes and to the linking of the actin cytoskeleton to the extracellular matrix. Its expression is thought to be restricted to striated muscle. Mutations in this gene result in type 2D autosomal recessive limb-girdle muscular dystrophy. Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Oct 2008],disease:Defects in SGCA are the cause of limb-girdle muscular dystrophy type 2D (LGMD2D) [MIM:608099]; also known as Duchenne-like muscular dystrophy autosomal recessive type 2 or severe childhood autosomal recessive muscular dystrophy (SCARMD). LGMD2D is an autosomal recessive degenerative myopathy characterized by progressive muscle wasting from early childhood with loss of independent ambulation by teenage years. Muscle biopsy shows necrosis, decreased immunostaining for alpha sarcoglycan, and adhalin deficiency. The phenotype is less severe than LGMD2C.,function:Component of the sarcoglycan complex, a subcomplex of the dystrophin-glycoprotein complex which forms a link between the F-actin cytoskeleton and the extracellular matrix.,online information:SGCA mutations in LGMD2D,similarity:Belongs to the sarcoglycan alpha/epsilon family.,subunit:Interacts with the syntrophin SNTA1. Cross-link to form 2 major subcomplexes: one consisting of SGCB, SGCD and SGCG and the other consisting of SGCB and SGCD. The association between SGCB and SGCG is particularly strong while SGCA is loosely associated with the other sarcoglycans.,tissue specificity:Most strongly expressed in skeletal muscle. Also expressed in cardiac muscle and, at much lower levels, in lung. In the fetus, most abundant in cardiac muscle and, at lower levels, in lung. Also detected in liver and kidney. Not expressed in brain.,