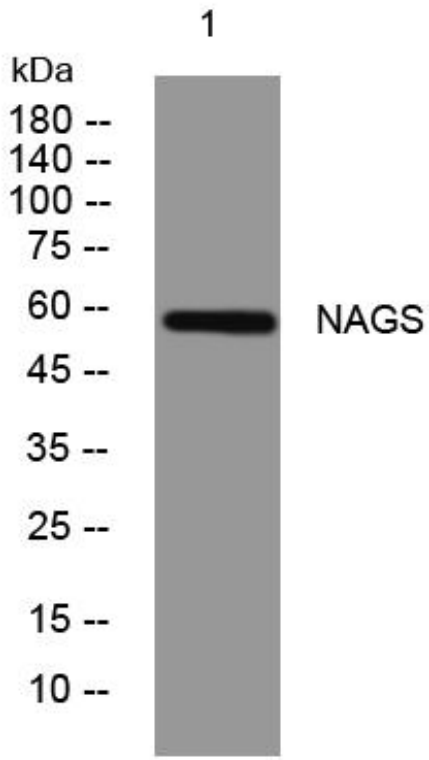
NAGS mouse mAb

Catalog No	BYmab-11407
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	NAGS
Protein Name	NAGS
Immunogen	Synthesized peptide derived from human NAGS AA range: 361-411
Specificity	This antibody detects endogenous levels of NAGS at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Mitochondrion matrix .
Tissue Specificity	Highly expressed in the adult liver, kidney and small intestine. Weakly expressed in the fetal liver, lung, pancreas, placenta, heart and brain tissue.
Function	catalytic activity:Acetyl-CoA + L-glutamate = CoA + N-acetyl-L-glutamate.,disease:Defects in NAGS are the cause of N-acetylglutamate synthase deficiency (NAGSD) [MIM:237310]. NAGSD is a rare autosomal recessively inherited metabolic disorder leading to severe neonatal or late onset hyperammonemia without increased excretion of orotic acid. Clinical symptoms are somnolence, tachypnea, feeding difficulties, a severe neurologic presentation characterized by uncontrollable movements, developmental delay, visual impairment, failure to thrive and hyperammonemia precipitated by the introduction of high-protein diet or febrile illness.,enzyme regulation:Increased by L-arginine.,function:Plays a role in the regulation of ureagenesis by producing variable amounts of N-acetylglutamate (NAG), thus modulating carbamoylphosphate synthase I (CPSI) activity.,online

	information:N-acetylglutamate synthase
Background	The N-acetylglutamate synthase gene encodes a mitochondrial enzyme that catalyzes the formation of N-acetylglutamate (NAG) from glutamate and acetyl coenzyme-A. NAG is a cofactor of carbamyl phosphate synthetase I (CPSI), the first enzyme of the urea cycle in mammals. This gene may regulate ureagenesis by altering NAG availability and, thereby, CPSI activity. Deficiencies in N-acetylglutamate synthase have been associated with hyperammonemia. [provided by RefSeq, Jul 2008],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using NAGS mouse mAb