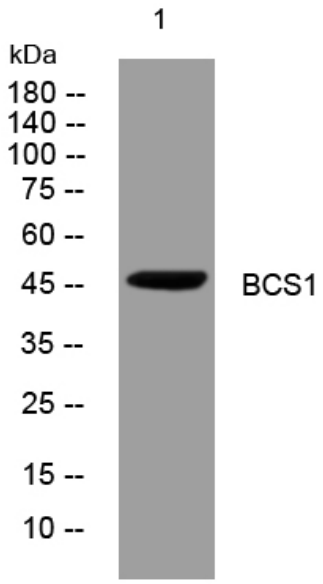
BCS1 rabbit pAb

Catalog No	BYab-08445
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	BCS1L BCS1
Protein Name	BCS1
Immunogen	Synthesized peptide derived from human BCS1 AA range: 29-79
Specificity	This antibody detects endogenous levels of BCS1 at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using 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 inner membrane ; Single-pass membrane protein .
Tissue Specificity	Ubiquitous.
Function	disease:Defects in BCS1L are a cause of mitochondrial complex III deficiency (CIII deficiency) [MIM:124000]. CIII deficiency is characterized by congenital lactic acidosis. Patients had severe failure to thrive, liver dysfunction and renal tubulopathy.,disease:Defects in BCS1L are the cause of Bjoernstad syndrome (BJS) [MIM:262000]. BJS is an autosomal recessive condition characterized by sensorineural hearing loss and pili torti. The hearing loss in BJS is congenital and of variable severity. Pili torti (twisted hairs), a condition in which the hair shafts are flattened at irregular intervals and twisted 180 degrees from the normal axis, making the hair extremely brittle, is usually recognized early in childhood.,disease:Defects in BCS1L are the cause of GRACILE syndrome [MIM:603358]. GRACILE stands for 'growth retardation, aminoaciduria, cholestasis, iron overload, lactic acidosis, and

Background	This gene encodes a homolog of the S. cerevisiae bcs1 protein which is involved in the assembly of complex III of the mitochondrial respiratory chain. The encoded protein does not contain a mitochondrial targeting sequence but experimental studies confirm that it is imported into mitochondria. Mutations in this gene are associated with mitochondrial complex III deficiency and the GRACILE syndrome. Several alternatively spliced transcripts encoding two different isoforms have been described. [provided by RefSeq, Jan 2016],
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 lysates from 3T3 cells, primary antibody was diluted at 1:1000, 4°over night