SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

Rabbit Anti-BFSP2 antibody

SL11015R

Product Name:	BFSP2
Chinese Name:	晶状体蛋白2抗体
Alias:	49 kDa cytoskeletal protein; Beaded filament protein CP49; Beaded filament structural protein 2; Beaded filament structural protein 2, phakinin; Bfps2, Cytoskeletal protein, 49 kD; BFSP2; BFSP2_HUMAN; CP47; CP49; Lens fiber cell beaded filament protein CP 47; Lens fiber cell beaded filament protein CP 49; Lens intermediate filament-like light; LIFL-L; Phakinin; PHAKOSIN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	46kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human BFSP2/Phakinin:181-280/415
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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
Product Detail:	Phakinin is a membrane-associated and cytoskeletal intermediate filament (IF) protein specific to the eye lens. IFs are cytoskeletal structures that typically contain a head, rod and tail domain. Unlike most IFs, Phakinin completely lacks the C-terminal tail domain

	thus contributing to the unique structure of the beaded filament that is specific to the lens. Phakinin is required for the assembly of beaded filaments and cytoskeletal networks that are important for the long-term maintenance of optical properties and transparency of the lens. Phakinin copolymerizes with Filensin, another IF protein, to form the 10-nm filamentous structures of the beaded filaments. Phakinin is also capable of self-assembling into filament-like structures that form thicker bundles. Mutations in the gene encoding Phakinin can result in lens cataract. **Function:** Involved in stabilization of lens fiber cell cytoskeleton. **Subunit:** Associates with BFSP1. Interacts with LGSN. **Subcellular Location:** Membrane. Cytoplasm. Cytoplasm, cytoskeleton. Membrane- and cytoskeleton-associated. **Tissue Specificity:** Lens. **DISEASE:** Defects in BFSP2 are the cause of cataract autosomal dominant BFSP2-related (ADC-BFSP2); also known as cataract autosomal dominant multiple types 1. Cataract is an opacification of the crystalline lens of the eye that frequently results in visual impairment or blindness. Opacities vary in morphology, are often confined to a portion of the lens, and may be static or progressive. In general, the more posteriorly located and dense an opacity, the greater the impact on visual function. Cataract autosomal dominant BFSP2-related is characterized by a variable phenotype that may or may not be consistent within a family. The opacities can be nuclear, sutural, stellate cortical, lamellar, cortical, nuclear embryonic, Y-sutural, punctate cortical, congenital or with juvenile- and adult-onset. **Similarity:** Belongs to the intermediate filament family. **SWISS:** Q13515 **Gene ID:** 8419 **Database links:** Entrez Gene: 8419 Human

	Omim: 603212 Human SwissProt: Q13515 Human Unigene: 659862 Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.