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Rabbit Anti-SIX1 antibody

SL17504R

Product Name:	SIX1
Chinese Name:	同源盒蛋白SIX1抗体
Alias:	BOS3; DFNA23; Homeobox protein SIX1; OTTHUMP00000179042; Sine oculis homeobox homolog 1; SIX homeobox 1; SIX1; SIX1_HUMAN; TIP39.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Rabbit,Sheep,
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:	32kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human SIX1:151-250/284
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:	The protein encoded by this gene is a homeobox protein that is similar to the Drosophila 'sine oculis' gene product. This gene is found in a cluster of related genes on chromosome 14 and is thought to be involved in limb development. Defects in this gene are a cause of autosomal dominant deafness type 23 (DFNA23) and branchiootic syndrome type 3 (BOS3). [provided by RefSeq, Jul 2008]

Function:
May be involved in limb tendon and ligament development.

Subcellular Location:
Nucleus.

Tissue Specificity:
Specifically expressed in skeletal muscle.

DISEASE:
Defects in SIX1 are the cause of deafness autosomal dominant type 23 (DFNA23) [MIM:605192]. A form of non-syndromic deafness characterized by prelingual, bilateral, symmetric hearing loss with a conductive component present in some but not all patients.
Defects in SIX1 are the cause of branchiootic syndrome type 3 (BOS3) [MIM:608389]. BOS3 is a syndrome characterized by usually bilateral branchial cleft fistulas or cysts, sensorineural and/or conductive hearing loss, pre-auricular pits, and structural defects of the outer, middle or inner ear. Otic defects include malformed and hypoplastic pinnae, a narrowed external ear canal, bulbous internal auditory canal, stapes fixation, malformed and hypoplastic cochlea. Branchial and otic anomalies are as those seen in individuals with the branchiootorenal syndrome. However, renal anomalies are absent in branchiootic syndrome patients.
Note=Defects in SIX1 could be a cause of branchiootorenal syndrome (BOR). BOR is an autosomal dominant disorder manifested by various combinations of preauricular pits, branchial fistulae or cysts, lacrimal duct stenosis, hearing loss, structural defects of the outer, middle, or inner ear, and renal dysplasia. Associated defects include asthenic habitus, long narrow facies, constricted palate, deep overbite, and myopia. Hearing loss may be due to mondini type cochlear defect and stapes fixation. Penetrance of BOR syndrome is high, although expressivity can be extremely variable.

Similarity:
Belongs to the SIX/Sine oculis homeobox family.
Contains 1 homeobox DNA-binding domain.

SWISS:
Q15475

Gene ID:
6495

Database links:

Entrez Gene: 6495 Human

Entrez Gene: 20471 Mouse

	Entrez Gene: 114634 Rat Omim: 601205 Human SwissProt: Q15475 Human SwissProt: Q62231 Mouse SwissProt: Q8BSP4 Mouse Unigene: 54416 Human Unigene: 713114 Human Unigene: 4645 Mouse Unigene: 23396 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.