CRX 抗体

产品货号： mlR3798

英文名称： CRX1

中文名称： CRX 抗体

别　　名： Crx; Crx-1; Crx 1; CRD; LCA7; CRX_HUMAN.

研究领域： 免疫学　信号转导　转录调节因子

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Dog, Pig, Cow, Horse, Rabbit,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 32kDa

细胞定位： 细胞核

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human CRX:51-150/299

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍 background:

The protein encoded by this gene is a photoreceptor-specific transcription factor which plays a role in the differentiation of photoreceptor cells. This homeodomain protein is necessary for the maintenance of normal

cone and rod function. Mutations in this gene are associated with photoreceptor degeneration, Leber congenital amaurosis type III and the autosomal dominant cone-rod dystrophy 2. Several alternatively spliced transcript variants of this gene have been described, but the full-length nature of some variants has not been determined. [provided by RefSeq, Jul 2008].

Function:

Binds and transactivates the sequence 5'-TAATC[CA]-3' which is found upstream of several photoreceptor-specific genes, including the opsin genes. Acts synergistically with other transcription factors, e.g. NRL and RX, to regulate photoreceptor cell-specific gene transcription. Essential for the maintenance of mammalian photoreceptors.

Subunit:

Interacts with SCA7. Interacts with RAX2. Interacts (via the homeobox) with NRL (via the leucine-zipper domain). Interacts with PDC.

Subcellular Location:

Nucleus.

Tissue Specificity:

Retina.

DISEASE:

Defects in CRX are the cause of Leber congenital amaurosis type 7 (LCA7) [MIM:613829]. LCA designates a clinically and genetically heterogeneous group of childhood retinal degenerations, generally inherited in an autosomal recessive manner. Affected infants have little or no retinal photoreceptor function as tested by electroretinography. LCA represents the most common genetic cause of congenital visual impairment in infants and children. [DISEASE] Defects in CRX are the cause of cone-rod dystrophy type 2 (CORD2) [MIM:120970]; also known as cone-rod retinal dystrophy 2 (CRD2). CORDs are inherited retinal dystrophies belonging to the group of

pigmentary retinopathies. CORDs are characterized by retinal pigment deposits visible on fundus examination, predominantly in the macular region, and initial loss of cone photoreceptors followed by rod degeneration. This leads to decreased visual acuity and sensitivity in the central visual field, followed by loss of peripheral vision. Severe loss of vision occurs earlier than in retinitis pigmentosa.

Defects in CRX are a cause of retinitis pigmentosa (RP) [MIM:268000]. RP leads to degeneration of retinal photoreceptor cells. Patients typically have night vision blindness and loss of midperipheral visual field. As their condition progresses, they lose their far peripheral visual field and eventually central vision as well.

Similarity:

Belongs to the paired homeobox family. Contains 1 homeobox DNA-binding domain.

SWISS:

O43186

Gene ID:

1406

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片