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Rabbit Anti-NR2E3 antibody

SL11572R

Product Name:	NR2E3
Chinese Name:	核受体蛋白NR2E3抗体
Alias:	ESCS; ESCS; NR2 E3; Nr2e3; NR2E3_HUMAN; Nuclear receptor subfamily 2 group E member 3; Photoreceptor specific nuclear receptor; Photoreceptor-specific nuclear receptor antibody PNR; Rd 7; rd7; Retina specific nuclear receptor; Retina-specific nuclear receptor; Retinal degeneration 7; Retinal degeneration 7; RNR; RP37.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,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:	45kDa
Cellular localization:	The nucleus
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human NR2E3:1-100/410
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:	Photoreceptor-specific nuclear receptor, also known as NR2E3 or PNR, belongs to a large family of nuclear hormone receptor transcription factors. The proteins belonging to this family are characterized by discrete domains functioning in DNA and ligand binding. NR2E3 has a role in regulating the signaling pathway elemental to the

	photoreceptor cell function and in regulating pathways involved in embryonic development. NR2E3 is an eye specific nuclear protein found in the outer nuclear layer of the adult retina (where the nuclei of cone and rod photoreceptors are located). Defects in this gene encoding for the protein, which localizes to chromosome 15q22.32, cause enhanced S cone syndrome. **Function:** Orphan nuclear receptor of retinal photoreceptor cells. Transcriptional factor that is an activator of rod development and repressor of cone development. Binds the promoter region of a number of rod- and cone-specific genes, including rhodopsin, M-and S-opsin and rod-specific phosphodiesterase beta subunit. Enhances rhodopsin expression. Represses M- and S-cone opsin expression. **Subunit:** Interacts with PIAS3; the interaction sumoylates NR2E3 and promotes repression of cone-specific gene transcription and activation of rod-specific genes (By similarity). Component of a complex that includes NR2E3, PIAS3, NRL, CRX and/or NR1D1. Binds NR1D1. Binds direcly in the complex with CRX, PIAS3 and NR1D1 (By similarity). Interacts (via the DNA-binding domain) with CRX (via its DNA binding domain); the interaction represses S- and M-cone opsin expression. **Subcellular Location:** Nucleus. **Tissue Specificity:** Eye specific; found solely in the outer nuclear layer of the adult neurosensory retina, where the nuclei of cone and rod photoreceptors reside. **Post-translational modifications:** Di- and tri-sumoylated in developing retina. PIAS3-mediated sumoylation promotes repression of cone-specific gene expression and activation of rod-specific genes. Sumoylation on Lys-185 appears to be the main site. **DISEASE:** Defects in NR2E3 are a cause of enhanced S cone syndrome (ESCS) [MIM:268100]. ESCS is an autosomal recessive retinopathy in which patients have increased sensitivity to blue light; perception of blue light is mediated by what is normally the least populous cone photoreceptor subtype, the S (short wavelength, blue) cones. ESCS is also associated with visual loss, with night blindness occurring from early in life, varying degrees of L (long, red)- and M (middle, green)-cone vision, and retinal degeneration. Defects in NR2E3 are the cause of retinitis pigmentosa type 37 (RP37) [MIM:611131]. 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. RP37 inheritance is autosomal dominant.

	Similarity: Belongs to the nuclear hormone receptor family. NR2 subfamily. Contains 1 nuclear receptor DNA-binding domain. **SWISS:** Q9Y5X4 **Gene ID:** 10002 **Database links:** Entrez Gene: 10002Human Omim: 604485Human SwissProt: Q9Y5X4Human Unigene: 187354Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.