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Product datasheet

Anti-Bestrophin/BEST1 antibody ab14927

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製品の概要

製品名	Anti-Bestrophin/BEST1 antibody
製品の詳細	Rabbit polyclonal to Bestrophin/BEST1
由来種	Rabbit
アプリケーション	**適用あり:** ICC, IP, ICC/IF, WB
種交差性	**交差種:** Mouse, Rat, Cow, Human **交差が予測される動物種:** Cynomolgus monkey
免疫原	Synthetic peptide corresponding to Human Bestrophin/BEST1 aa 1-100. Database link: O76090 Run BLAST with Run BLAST with
特記事項	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
バッファー	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: Tris glycine, 0.5% BSA, 30% Glycerol
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、** 次のテスト済みアプリケーションにおけるab14927の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
ICC	★★★★★ (1)	Use at an assay dependent concentration.
IP	★★★★☆ (1)	Use at an assay dependent concentration.
ICC/IF	★★★★☆ (1)	Use at an assay dependent concentration.
WB	★★★★☆ (2)	Use at an assay dependent concentration.

ターゲット情報

機能 Forms calcium-sensitive chloride channels. Highly permeable to bicarbonate.

組織特異性 Predominantly expressed in the basolateral membrane of the retinal pigment epithelium.

関連疾患 Defects in BEST1 are the cause of vitelliform macular dystrophy type 2 (VMD2) [MIM:153700]; also known as Best macular dystrophy (BMD). VMD2 is an autosomal dominant form of macular degeneration that usually begins in childhood or adolescence. VMD2 is characterized by typical 'egg-yolk' macular lesions due to abnormal accumulation of lipofuscin within and beneath the retinal pigment epithelium cells. Progression of the disease leads to destruction of the retinal pigment epithelium and vision loss.
Defects in BEST1 are the cause of retinitis pigmentosa type 50 (RP50) [MIM:613194]. A retinal dystrophy belonging to the group of pigmentary retinopathies. RP is characterized by retinal pigment deposits visible on fundus examination and primary loss of rod photoreceptor cells followed by secondary loss of cone photoreceptors. 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.
Defects in BEST1 are a cause of adult-onset vitelliform macular dystrophy (AVMD) [MIM:608161]. AVMD is a rare autosomal dominant disorder with incomplete penetrance and highly variable expression. Patients usually become symptomatic in the fourth or fifth decade of life with a protracted disease of decreased visual acuity.
Defects in BEST1 are the cause of bestrophinopathy autosomal recessive (ARB) [MIM:611809]. A retinopathy characterized by central visual loss, an absent electro-oculogram light rise, and a reduced electroretinogram.
Defects in BEST1 are the cause of vitreoretinochoroidopathy autosomal dominant (ADVIRC) [MIM:193220]. A disorder characterized by vitreoretinochoroidal dystrophy. The clinical presentation is variable and may be associated with cataract, nanophthalmos, microcornea, shallow anterior chamber, and glaucoma.

配列類似性 Belongs to the bestrophin family.

翻訳後修飾 Phosphorylated by PP2A.

細胞内局在 Cell membrane. Basolateral cell membrane.

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