abcam

Product datasheet

Anti-PAX3 antibody [6288D4a] ab53571

1 References 1 图像

概述

产品名称	Anti-PAX3抗体[6288D4a]
描述	**小鼠单克隆抗体**[6288D4a] to PAX3
宿主	Mouse
经测试应用	**适用于:** WB
种属反应性	**与反应:** Recombinant fragment
免疫原	Recombinant fragment corresponding to Human PAX3.
常规说明	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

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
存储溶液	pH: 7.40 Preservative: 0.05% Sodium azide Constituents: 1% BSA, 0.03% Tripotassium orthophosphate, 0.812% Sodium chloride, 0.1312% Sodium phosphate, 0.0225% Potassium chloride, PBS
纯度	Protein G purified
纯化说明	ab53571 was purified using protein G column chromatography from culture supernatant of hybridoma cultured in a medium containing bovine IgG-depleted (approximately 95%) fetal bovine serum and filtered through a 0.22µm membrane.
克隆	单克隆
克隆编号	6288D4a
同种型	IgG1

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab53571于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
WB		Use at an assay dependent concentration. Detects a band of approximately 40 kDa (predicted molecular weight: 53 kDa).

靶标

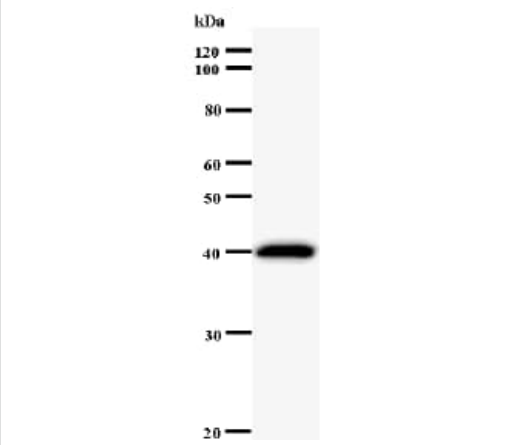
功能 Probable transcription factor associated with development of alveolar rhabdomyosarcoma.

疾病相关 Defects in PAX3 are the cause of Waardenburg syndrome type 1 (WS1) [MIM:193500]. WS1 is an autosomal dominant disorder characterized by wide bridge of nose owing to lateral displacement of the inner canthus of each eye (dystopia canthorum), pigmentary disturbances such as frontal white blaze of hair, heterochromia of irides, white eyelashes, leukoderma and sensorineural deafness. The syndrome shows variable clinical expression and some affected individuals do not manifest hearing impairment.
Defects in PAX3 are the cause of Waardenburg syndrome type 3 (WS3) [MIM:148820]; also known as Klein-Waardenburg syndrome or Waardenburg syndrome with upper limb anomalies or white forelock with malformations. WS3 is a very rare autosomal dominant disorder, which shares many of the characteristics of WS1. Patients additionally present with musculoskeletal abnormalities.
Defects in PAX3 are the cause of craniofacial-deafness-hand syndrome (CDHS) [MIM:122880]. CDHS is thought to be an autosomal dominant disease which comprises absence or hypoplasia of the nasal bones, hypoplastic maxilla, small and short nose with thin nares, limited movement of the wrist, short palpebral fissures, ulnar deviation of the fingers, hypertelorism and profound sensory-neural deafness.
Defects in PAX3 are a cause of rhabdomyosarcoma type 2 (RMS2) [MIM:268220]. It is a form of rhabdomyosarcoma, a highly malignant tumor of striated muscle derived from primitive mesenchimal cells and exhibiting differentiation along rhabdomyoblastic lines.
Rhabdomyosarcoma is one of the most frequently occurring soft tissue sarcomas and the most common in children. It occurs in four forms: alveolar, pleomorphic, embryonal and botryoidal rhabdomyosarcomas. Note=A chromosomal aberration involving PAX3 is found in rhabdomyosarcoma. Translocation (2;13)(q35;q14) with FOXO1. The resulting protein is a transcriptional activator.
Note=A chromosomal aberration involving PAX3 is a cause of rhabdomyosarcoma. Translocation t(2;2)(q35;p23) with NCOA1 generates the NCOA1-PAX3 oncogene consisting of the N-terminus part of PAX3 and the C-terminus part of NCOA1. The fusion protein acts as a transcriptional activator. Rhabdomyosarcoma is the most common soft tissue carcinoma in childhood, representing 5-8% of all malignancies in children.

序列相似性 Belongs to the paired homeobox family.
Contains 1 homeobox DNA-binding domain.
Contains 1 paired domain.

细胞定位 Nucleus.

图片



Western blot - Anti-PAX3 antibody [6288D4a] (ab53571)

Anti-PAX3 antibody [6288D4a] (ab53571) + immunised recombinant protein

Predicted band size: 53 kDa
Observed band size: 40 kDa

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