# abcam

# Product datasheet

# Anti-Nance-Horan Syndrome Protein antibody ab122177

# 画像数 2

#### 製品の概要

製品名 Anti-Nance-Horan Syndrome Protein antibody

製品の詳細 Rabbit polyclonal to Nance-Horan Syndrome Protein

由来種 Rabbit

アプリケーション 適用あり: ICC/IF, IHC-P

種交差性 交差種: Human

免疫原 antigen sequence, corresponding to amino acids 1139-1281 of Human Nance-Horan Syndrome

Protein.

ポジティブ・コントロール Human liver tissue.

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 Liquic

保存方法 Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.

**バッファー** pH: 7.20

Preservative: 0.02% Sodium azide

Constituents: 40% Glycerol (glycerin, glycerine), 59% PBS

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

#### 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/IF		Use a concentration of 1 - 4 µg/ml. Recommend PFA Fixation and Triton X-100 treatment
IHC-P		1/20 - 1/50. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

#### ターゲット情報

関連疾患

機能 Unknown. May have a key functions in the regulation of eye, tooth, brain and craniofacial

development.

**組織特異性**Detected at low levels in all tissues analyzed. Detected in fetal and adult brain, lens, retinal pigment epithelium, placenta, lymphocytes and fibroblasts. Levels in retinal pigment epithelium,

placenta, lymphocytes, and fibroblasts are very low. Expressed also in kidney, lung and thymus.

Defects in NHS are the cause of Nance-Horan syndrome (NHS) [MIM:302350]; also known as cataract-dental syndrome. NHS is a rare X-linked disorder characterized by congenital cataracts, dental anomalies, dysmorphic features, and, in some cases, mental retardation. Distinctive dental

anomalies are seen in affected males, including supernumerary incisors and crown shaped permanent teeth. Characteristic facial features are anteverted pinnae, long face, and prominent nasal bridge and nose. Carrier females display milder variable symptoms of disease with lens opacities often involving the posterior Y sutures, and on occasion dental anomalies and the

characteristic facial features described.

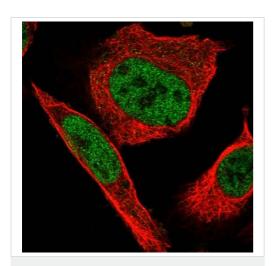
Defects in NHS are the cause of cataract congenital X-linked (CXN) [MIM:302200]. A X-linked form of cataract, manifesting as a congenital nuclear opacity with severe visual impairment in affected males. Heterozygous females have suture cataracts and only slight reduction in vision. Note=Caused by copy number variations predicted to result in altered transcriptional regulation of the NHS gene: a 0.8 Mb segmental duplication-triplication encompassing the NHS, SCML1 and

RAI2 genes, and an 4.8 kb intragenic deletion in NHS intron 1.

配列類似性 Belongs to the NHS family.

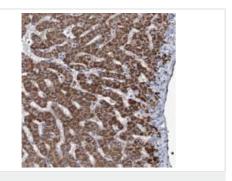
細胞内局在 Nucleus.

#### 画像



Immunocytochemistry/ Immunofluorescence - Anti-Nance-Horan Syndrome Protein antibody (ab122177)

Immunofluorescent staining of Human cell line U-2 OS shows positivity in nucleus but not nucleoli. Recommended concentration of ab122177 1-4  $\mu$ g/ml. Cells treated with PFA/Triton X-100.



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-Nance-Horan Syndrome
Protein antibody (ab122177)

ab122177 at 1/35 dilution staining Nance-Horan Syndrome Protein in Paraffin-embedded Human Liver tissue by Immunohistochemistry.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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