

# Anti-Nance-Horan Syndrome Protein antibody ab122177

## 2 图像

### 概述

<b>产品名称</b>	Anti-Nance-Horan Syndrome蛋白抗体
<b>描述</b>	兔多克隆抗体to Nance-Horan Syndrome蛋白
<b>宿主</b>	Rabbit
<b>经测试应用</b>	<b>适用于:</b> ICC/IF, IHC-P
<b>种属反应性</b>	<b>与反应:</b> Human
<b>免疫原</b>	antigen sequence, corresponding to amino acids 1139-1281 of Human Nance-Horan Syndrome Protein.
<b>阳性对照</b>	Human liver tissue.
<b>常规说明</b>	<p>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.</p> <p>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&amp;As</p>

### 性能

<b>形式</b>	Liquid
<b>存放说明</b>	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
<b>存储溶液</b>	pH: 7.20 Preservative: 0.02% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), 59% PBS
<b>纯度</b>	Immunogen affinity purified
<b>克隆</b>	多克隆
<b>同种型</b>	IgG

### 应用

The Abpromise guarantee

**Abpromise™**承诺保证使用ab122177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
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.

## 靶标

### 功能

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

### 组织特异性

Detected at low levels in all tissues analyzed. Detected in fetal and adult brain, lens, retina, 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.

### 疾病相关

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.

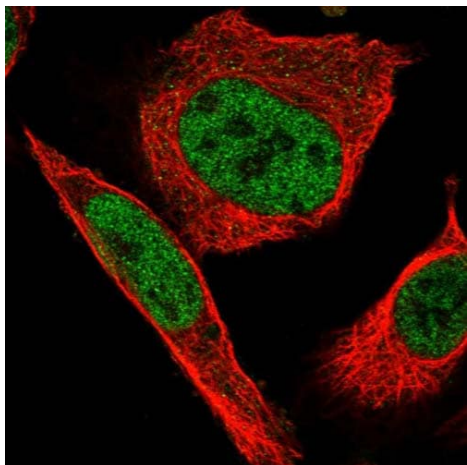
### 序列相似性

Belongs to the NHS family.

### 细胞定位

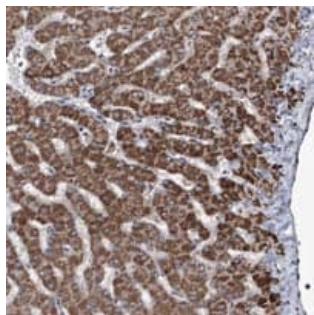
Nucleus.

## 图片



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

Immunocytochemistry/ Immunofluorescence - 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.

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

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

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