核仁蛋白 5A 抗体

产品货号： mlR19311

英文名称： NOL5A/NOP56

中文名称： 核仁蛋白 5A 抗体

别　　名： NOL5A; NOP56; NOP56 ribonucleoprotein; NOP56 ribonucleoprotein homolog (yeast); NOP56 ribonucleoprotein homolog; Nucleolar protein 56; Nucleolar protein 5A (56kD with KKE/D repeat); Nucleolar protein 5A; Nucleolar protein Nop56; NOP56_HUMAN; RP4-686C3.1; SCA36.

研究领域： 细胞生物　染色质和核信号　转录调节因子　表观遗传学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Pig,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 66kDa

细胞定位： 细胞核

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human NOL5A/NOP56:301-400/594

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： NOL5A is similar in sequence to Nop56p, a yeast nucleolar protein that is part of a complex with the nucleolar proteins Nop58p and fibrillarin. Like Nop56p, NOL5A is found in the nucleolus and is though to be required for 60S ribosomal subunit biogenesis. Multiple transcript variants encoding several isoforms have been identified for this gene, but the full-length nature of most of them is not yet determined.

Function:

Involved in the early to middle stages of 60S ribosomal subunit biogenesis. Core component of box C/D small nucleolar ribonucleoprotein (snoRNP) particles. Required for the biogenesis of box C/D snoRNAs such U3, U8 and U14 snoRNAs.

Subcellular Location:

Nuclear; nucleolus

DISEASE:

Spinocerebellar ataxia 36 (SCA36) [MIM:614153]: A form of spinocerebellar ataxia, a clinically and genetically heterogeneous group of cerebellar disorders. Patients show progressive incoordination of gait and often poor coordination of hands, speech and eye movements, due to degeneration of the cerebellum with variable

involvement of the brainstem and spinal cord. SCA36 is characterized by complicated clinical features, with ataxia as the first symptom, followed by characteristic late-onset involvement of the motor neuron system. Ataxic symptoms, such as gait and truncal instability, ataxic dysarthria, and uncoordinated limbs, start in late forties to fifties. Characteristically, affected individuals exhibit tongue atrophy with fasciculation. Progression of motor neuron involvement is typically limited to the tongue and main proximal skeletal muscles in both upper and lower extremities.

Note: The disease is caused by mutations affecting the gene represented in this entry. Caused by large hexanucleotide CGCCTG repeat expansions within intron 1. These expansions induce RNA foci and sequester the RNA-binding protein SRSF2. In addition, the transcription of MIR1292, a microRNA gene located just 19 bp 3' of the GGCCTG repeat, is significantly decreased.

Similarity:

Belongs to the NOP5/NOP56 family. {ECO:0000305}.

Contains 1 Nop domain.

SWISS:

O00567

Gene ID:

10528

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.