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DATA SHEET

Mouse Anti-PolyQ/Polyglutamine-Expansion Diseases Marker [5TF1-1C2]: MC0046

Intended Use: For Research Use Only

Description: Polyglutamine tract are portions of a protein consisting of a sequence of several glutamine unit. Several inheritable neurodegenerative disorders, so-called polyglutamine diseases, occur if a mutation causes a polyglutamine tract in a specific gene to become too long. Important examples of polyglutamine diseases include Huntington's disease, dentatorubralpallidoluysian atrophy (DRPLA), spinobulbar muscular atrophy (SBMA) and types of spinocerebellar ataxia (SCA). In these diseases, the pathogenic alleles usually contain 39 or more consecutive glutamine repeats. Higher repeat numbers lead to lower ages of onset. Patients with 40-60 glutamine repeats normally develop disease as adults, whereas patients with more than 60 repeats develop a juvenile onset disease. Each polyglutamine expansion disorder displays characteristic pathology, with neuronal loss evident in specific regions of the brain. It is believed that cells cannot properly dispose of proteins with overlong polyglutamine tracts, which over time leads to damage in nerve cells.

Specifications

Clone:	5TF1-1C2
Source:	Mouse
Isotype:	IgG1k
Reactivity:	Human
Localization:	Nucleus
Formulation:	Antibody in PBS pH7.4, containing BSA and ≤ 0.09% sodium azide (NaN3)
Storage:	Store at 2°- 8°C
Applications:	IHC, ELISA, ICC, IP, WB

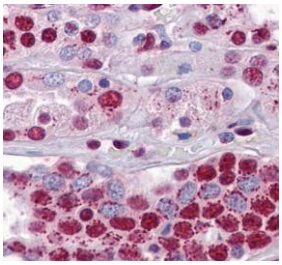
Package:

Description	Catalog No.	Size
PolyQ/Polyglutamine-Expansion Diseases Marker Concentrated	MC0046	1 ml

IHC Procedure*

Positive Control Tissue:	Small intestine
Concentrated Dilution:	50-500
Pretreatment:	Citrate pH6.0 or EDTA pH8.0, 15 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C
Incubation Time and Temp:	30-60 minutes @ RT
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human kidney stained with anti-PolyQ using AEC

References:

1. Inactivation of PNKP by mutant ATXN3 triggers apoptosis by activating the DNA damage-response pathway in SCA3. Gao, R; Liu, Y; Silva-Fernandes, A; et al. PLoS genetics 11 e1004834 , 2015.
2. Proteotoxic stress induces phosphorylation of p62/SQSTM1 by ULK1 to regulate selective autophagic clearance of protein aggregates. Lim, J; Lachenmayer, ML; et al. PLoS genetics 11 e1004987, 2015.
3. Development and characterization of 3-(benzylsulfonamido)benzamides as potent and selective SIRT2 inhibitors. Khanfar, MA; Quinti, L; et al. European journal of medicinal chemistry 76 414-26, 2014.

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