



Mjd, Machado-Joseph disease polyclonal antibody

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Box 1 | Basic Info

Cat. No.	ABP-PAB-10836
Animal ID	RB0321-0322
Host	Rabbit
Reactivity	Mouse
Format	Purified
Accession number	XM_127081
Amount	100µg

Alternative Name(s):

spinocerebellar ataxia 3 protein, autosomal dominant olivopontocerebellar ataxia 3 protein, ataxin 3, ATX3, MJD1, Sca3, 2210008M02Rik, josphin

Machado-Joseph disease is an autosomal dominant neurologic disorder, and is now known to be identical with spinocerebellar ataxia-3. Machado-Joseph disease protein (Mjd) contains a stretch of (CAG)_n repeats in the coding region, and the expansion of these repeats from the normal 13-36 to 68-79 is the cause of Machado-Joseph disease. There is a negative correlation between the age of onset and CAG repeat numbers. Live-cell imaging reveals divergent intracellular dynamics of polyglutamine disease proteins and supports a sequestration model of pathogenesis.

Buffers

Purified rabbit polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein G column and eluted out with both high and low pH buffers and neutralized immediately after elution then followed by dialysis against PBS.

Immunogen

KLH conjugated synthetic peptide comprised of amino acids 30 - 45 [PVELSSIAHQLDEEER] of the mouse Machado-Joseph disease (Mjd) protein.

Application

Tested by peptide-specific ELISA (1:1,000).

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C. Avoid repeated freeze-thaw cycles.

References:

1. Yoshida H, Yoshizawa T, Shibasaki F, Shoji S, Kanazawa I: Chemical chaperones reduce aggregate formation and cell death caused by the truncated Machado-Joseph disease gene product with an expanded polyglutamine stretch. *Neurobiol. Dis.* 10(2): 88-99 (2002).
2. Wang G, Sawai N, Kotliarova S, Kanazawa I, Nukina N: Ataxin-3, the MJD1 gene product, interacts with the two human homologs of yeast DNA repair protein RAD23, HHR23A and HHR23B. *Hum. Mol. Genet.* 9(12): 1795-1803 (2000).
3. Tait D, Riccio M, Sittler A, Scherzinger E, Santi S, Ognibene A, Maraldi NM, Lehrach H, Wanker EE: Ataxin-3 is transported into the nucleus and associates with the nuclear matrix. *Hum. Mol. Genet.* 7(6): 991-997 (1998).
4. Goto J, Watanabe M, Ichikawa Y, Yee SB, Ihara N, Endo K, Igarashi S, Takiyama Y, Gaspar C, Maciel P, Tsuji S, Rouleau GA, Kanazawa I: Machado-Joseph disease gene products carrying different carboxyl termini. *Neurosci. Res.* 28(4): 373-377 (1997). Review.
5. Paulson HL, Das SS, Crino PB, Perez MK, Patel SC, Gotsdiner D, Fischbeck KH, Pittman RN: Machado-Joseph disease gene product is a cytoplasmic protein widely expressed in brain. *Ann. Neurol.* 41(4): 453-462 (1997).
6. Ikeda H, Yamaguchi M, Sugai S, Aze Y, Narumiya S, Kakizuka A: Expanded polyglutamine in the Machado-Joseph disease protein induces cell death in vitro and in vivo. *Nat. Genet.* 13(2): 196-202 (1996).