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## 36-2331: Anti-Frataxin (Marker of Friedreich Ataxia) Monoclonal Antibody(Clone: FXN/2124)

Clonality: Monoclonal Clone Name: FXN/2124 Application: WB.IHC Reactivity: Human Gene: **FXN** 2395 Gene ID: **Uniprot ID:** Q16595

CyaY; d-FXN; FARR; Frataxin mature form; Frataxin (81-210); FRDA; Friedreich ataxia protein; **Alternative Name:** 

Fxn; i-FXN; m56-FXN; m78-FXN; m81-FXN; X25

Isotype: Mouse IgG2b, kappa

Recombinant fragment of human Frataxin (FXN) protein (around aa 57-210) (exact sequence is Immunogen Information:

proprietary)

## **Description**

Frataxin is a nuclear-encoded mitochondrial protein implicated in Friedreich's ataxia (FRDA), a human autosomal recessive neurodegenerative disease. Lack of Frataxin causes iron to accumulate in the mitochondrial matrix suggesting that Frataxin is involved in mitochondrial iron homeostasis and possibly in iron transport. Frataxin has an alpha-beta fold consisting of two helices flanking an antiparallel beta sheet.

## **Product Info**

Amount: 20 μg / 100 μg

200 µg/ml of Ab Purified from Bioreactor Concentrate by Protein A/G. Prepared in 10mM PBS with Content:

0.05% BSA & 0.05% azide. Also available WITHOUT BSA & azide at 1.0mg/ml.

Antibody with azide - store at 2 to 8°C. Antibody without azide - store at -20 to -80°C. Antibody is Storage condition:

stable for 24 months. Non-hazardous.

## **Application Note**

Western Blot (1-2ug/ml);Immunohistochemistry (Formalin-fixed) (1-2ug/ml for 30 min at RT)(Staining of formalin-fixed tissues requires heating tissue sections in 10mM Tris with 1mM EDTA, pH 9.0, for 45 min at 95&degC followed by cooling at RT for 20 minutes);

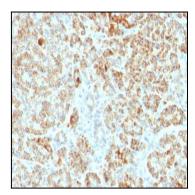


Fig. 1: Formalin-fixed, paraffin-embedded human Pancreas stained with Frataxin Mouse Monoclonal Antibody (FXN/2124).



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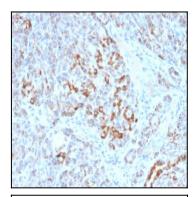


Fig. 2: Formalin-fixed, paraffin-embedded human Pancreas stained with Frataxin Mouse Monoclonal Antibody (FXN/2124).

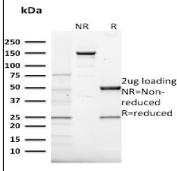


Fig. 3: SDS-PAGE Analysis Purified Frataxin Mouse Monoclonal Antibody (FXN/2124). Confirmation of Integrity and Purity of Antibody.

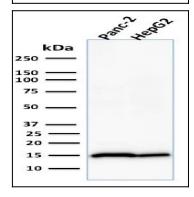


Fig. 4: Western Blot Analysis of Human Panc-2 and HepG2 cell lysate using Frataxin Mouse Monoclonal Antibody (FXN/2124).

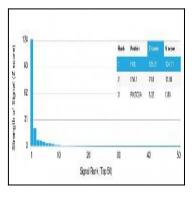


Fig. 5: Analysis of Protein Array containing more than 19,000 full-length human proteins using Frataxin Mouse Monoclonal Antibody (FXN/2124). Z- and S- Score: The Z-score represents the strength of a signal that a monoclonal antibody (MAb) (in combination with a fluorescently-tagged anti-IgG secondary antibody) produces when binding to a particular protein on the HuProtTM array. Z-scores are described in units of standard deviations (SD's) above the mean value of all signals generated on that array. If targets on HuProtTM are arranged in descending order of the Z-score, the S-score is the difference (also in units of SD's) between the Z-score. S-score therefore represents the relative target specificity of a MAb to its intended target. A MAb is considered to specific to its intended target, if the MAb has an S-score of at least 2.5. For example, if a MAb binds to protein X with a Z-score of 43 and to protein Y with a Z-score of 14, then the S-score for the binding of that MAb to protein X is equal to 29.