

## Anti-PDHA1 Antibody (1V504)

## Product Details

|               |              |
|---------------|--------------|
| Ig Type:      | Rabbit IgG   |
| Reactivity:   | Human        |
| Conjugation:  | Unconjugated |
| Clone:        | 1V504        |
| Purification: | Protein A    |

## Applications

|                    |                                                                                                                                                                                                                                                                                                                                                                                                                                                              |
|--------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Verified Activity: | Immunofluorescence staining of PDHA1 in HeLa cells. Cells were fixed with 4% PFA, permeabilized with 0.1% Triton X-100 in PBS, blocked with 10% serum, and incubated with rabbit anti-Human PDHA1 monoclonal antibody (dilution ratio 1:60) at 4°C overnight. Then cells were stained with the Alexa Fluor®488-conjugated Goat Anti-rabbit IgG secondary antibody (green) and counterstained with DAPI (blue). Positive staining was localized to Cytoplasm. |
| Application:       | ICC/IF                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
| Recommended        | ICC-IF: 1:20-1:100                                                                                                                                                                                                                                                                                                                                                                                                                                           |

## Properties

|                      |                                                                                                                            |
|----------------------|----------------------------------------------------------------------------------------------------------------------------|
| Stability & Storage: | Store at 2°C-8°C for 1 month. Store at -20°C or -80°C for 12 months. Avoid repeated freeze-thaw cycles. Preservative-Free. |
| Shipping:            | Shipping with blue ice.                                                                                                    |

## Antigen Details

|                  |                                          |
|------------------|------------------------------------------|
| Immunogen:       | Recombinant Protein: Human PDHA1 Protein |
| Antigen Species: | Human                                    |

## Research Background

PDHA1, also known as C54G1, is an alpha subunit of pyruvate dehydrogenase. Pyruvate dehydrogenase, together with dihydrolipoamide acetyltransferase and lipoamide dehydrogenase, composes the pyruvate dehydrogenase (PDH) complex. The PDH complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA and CO<sub>2</sub>, and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. PDHA1 plays a key role in the function of the PDH complex. Defects in PDHA1 can cause pyruvate dehydrogenase E1-alpha deficiency. Defects in PDHA1 also are the cause of X-linked Leigh syndrome (X-LS). X-LS is an early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in one or more areas of the central nervous system, including the brainstem, thalamus, basal ganglia, cerebellum, and spinal cord.

**Inhibitor · Natural Compounds · Compound Libraries · Recombinant Proteins**

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