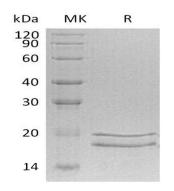
## Recombinant Human MCEE Protein (His Tag)

## Catalog Number: PDMH100013

Note: Centrifuge before opening to ensure complete recovery of vial contents.

Description	
Species	Human
Source	HEK293 Cells-derived Human MCEE protein Gln37-Ala176, with an C-terminal His
Calculated MW	16.0 kDa
Observed MW	18-20 kDa
Accession	Q96PE7
Bio-activity	Not validated for activity
Properties	
Purity	> 95% as determined by reducing SDS-PAGE.
Endotoxin	< 1.0 EU/mg of the protein as determined by the LAL method
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80
	°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of
	reconstituted samples are stable at $< -20^{\circ}$ C for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
Formulation	Lyophilized from a 0.2 $\mu$ m filtered solution in PBS with 5% Trehalose and 5%
	Mannitol.
Reconstitution	It is recommended that sterile water be added to the vial to prepare a stock solution of
	0.5 mg/mL. Concentration is measured by UV-Vis.

## Data



> 95 % as determined by reducing SDS-PAGE.

## Background

Methylmalonyl-CoA epimerase, mitochondrial (MCEE) is an enzyme which belongs to the glyoxalase I family. It converts (S)-methylmalonyl-CoA to the (R) form, catalyses the following chemical reaction: (R)-methylmalonyl-CoA (S)methylmalonyl-CoA. It plays an important role in the catabolism of fatty acids with odd-length carbon chains. This protein deficiency is an autosomal recessive inborn error of AA metabolism, involving valine, threonine, isoleucine and methionine. This organic aciduria can appear in the neonatal period with life-threatening metabolic acidosis, hyperammonemia, feeding difficulties, pancytopenia and coma.