

## Recombinant Human SDHA Protein (His Tag)

Catalog Number: PDEH100987

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

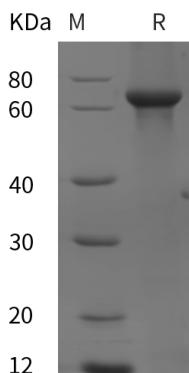
### Description

Species	Human
Source	E.coli-derived Human SDHA protein Ala43-Tyr664, with an N-terminal His & C-terminal His
Calculated MW	68.3 kDa
Observed MW	70 kDa
Accession	P31040
Bio-activity	Not validated for activity

### Properties

Purity	> 95% as determined by reducing SDS-PAGE.
Endotoxin	< 10 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°C for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
Formulation	Lyophilized from a 0.2 µm 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



SDS-PAGE analysis of Human SDHA proteins, 2 µg/lane of Recombinant Human SDHA proteins was resolved with SDS-PAGE under reducing conditions, showing bands at 70 kDa.

### Background

Flavoprotein (FP) subunit of succinate dehydrogenase (SDH) that is involved in complex II of the mitochondrial electron transport chain and is responsible for transferring electrons from succinate to ubiquinone. Defects in SDHA are a cause of mitochondrial complex II deficiency (MT-C2D). A disorder of the mitochondrial respiratory chain with heterogeneous clinical manifestations. Clinical features include psychomotor regression in infants, poor growth with lack of speech development, severe spastic quadriplegia, dystonia, progressive leukoencephalopathy, muscle weakness, exercise intolerance, cardiomyopathy. Some patients manifest Leigh syndrome or Kearns-Sayre syndrome.

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