Recombinant Human HSPD1/HSP60 Protein (His &GST Tag)

Catalog Number: PKSH031150

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

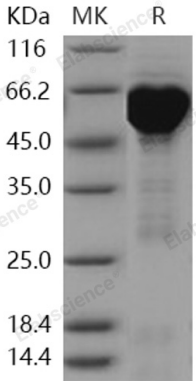
Description

Species	Human
Source	E.coli-derived Human HSPD1/HSP60 protein Leu 2-Phe 573, with an N-terminal His & GST
Calculated MW	88.7 kDa
Observed MW	52-65 kDa
Accession	NP_955472.1
Bio-activity	Not validated for activity

Properties

Purity	> 90 % as determined by reducing SDS-PAGE.
Endotoxin	Please contact us for more information.
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 sterile PBS, pH 7.4 Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
Reconstitution	Please refer to the printed manual for detailed information.

Data



> 90 % as determined by reducing SDS-PAGE.

Background

HSPD1, also known as HSP60, is a member of the chaperonin family. HSPD1 may function as a signaling molecule in the innate immune system. This protein is essential for the folding and assembly of newly imported proteins in the mitochondria. It may also prevent misfolding and promote the refolding and proper assembly of unfolded polypeptides generated under stress conditions in the mitochondrial matrix. HSPD1 gene is adjacent to a related family member and the region between the 2 genes functions as a bidirectional promoter. Several pseudogenes have been associated with this gene. Mutations associated with this gene cause autosomal recessive spastic paraplegia 13. Defects in HSPD1 are a cause of spastic paraplegia autosomal dominant type 13 (SPG13). Spastic paraplegia is a degenerative spinal cord disorder characterized by a slow, gradual, progressive weakness and spasticity of the lower limbs. Defects in HSPD1 are the cause of leukodystrophy hypomyelinating type 4 (HLD4); also called mitochondrial HSP60 chaperonopathy or MitCHAP-60 disease. HLD4 is a severe autosomal recessive hypomyelinating leukodystrophy. HSPD1 is cinically characterized by infantile-onset rotary nystagmus, progressive spastic paraplegia, neurologic regression, motor impairment, profound mental retardation. Death usually occurrs within the first two decades of life.

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