# Mouse HSPD1 / HSP60 Protein (His Tag)

Catalog Number: 50596-M07E



# **General Information**

# Gene Name Synonym:

60kDa; Hsp60

### **Protein Construction:**

A DNA sequence encoding the mouse HSP60 (NP\_034607.3) (Leu 2-Phe 573) was expressed, with a polyhistide tag at the N-terminus.

Source: Mouse

Expression Host: E. coli

**QC** Testing

Purity: > 95 % as determined by SDS-PAGE

**Endotoxin:** 

Please contact us for more information.

Stability:

Samples are stable for up to twelve months from date of receipt at -70 °C

Predicted N terminal: Met

### **Molecular Mass:**

The recombinant mouse HSP60 consisting of 583 amino acids and has a calculated molecular mass of 62.3 kDa. rmHSP60 migrates as an approximately 58 kDa band in SDS-PAGE under reducing conditions.

### Formulation:

Lyophilized from sterile PBS, pH 7.4

Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

# **Usage Guide**

### Storage:

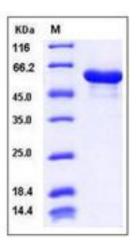
Store it under sterile conditions at  $-20\,^\circ\!\mathrm{C}$  to  $-80\,^\circ\!\mathrm{C}$  upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

## Reconstitution:

Detailed reconstitution instructions are sent along with the products.

### SDS-PAGE:



# **Protein Description**

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.

#### References

1.Hansen J J, et al. (2002) Hereditary spastic paraplegia SPG13 is associated with a mutation in the gene encoding the mitochondrial chaperonin Hsp60. Am J Hum Genet. 70: 1328-32. 2.Magen D, et al. (2008) Mitochondrial Hsp60 chaperonopathy causes an autosomal-recessive neurodegenerative disorder linked to brain hypomyelination and leukodystrophy. Am J Hum Genet. 83: 30-42. 3.Venner TJ, et al. (1990) Nucleotide sequences and novel structural features of human and Chinese hamster hsp60 (chaperonin) gene families. DNA Cell Biol. 9 (8): 545-52.

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