abcam

Product datasheet

Recombinant Human Myelin Protein Zero ab114281

画像数 1

製品の詳細

製品名	Recombinant Human Myelin Protein Zero
発現系	Wheat germ
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	MLRAPAPAPAMAPGAPSSSPSPILAVLLFSSLVLSPAQAIVV YTDREAHG AVGSRVTLHCSFWSSEWVSDDISFTWRYQPEGGRDAISIFHY AKGQPYID EVGTFKERIQWVGDPRWKDGSIVIHNLDYSDNGTFTCDVKNP PDIVGKTS QVTLYVFEKVPTRYGVVLGAVIGGVLGVVLLLLLLFYVVRYC WLRRQAAL QRRLSAMEKGKLHKPGKDASKRGRQTPVLYAMLDHSRSTKAV SEKKAKGL GESRKDKK
予測される分子量	54 kDa including tags
領域	1 to 258
製品の詳細	Recombinant Human Myelin Protein Zero

特性

Our **Abpromise guarantee** covers the use of **ab114281** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリケーション	ELISA
	SDS-PAGE
	Western blot
製品の状態	Liquid

前処理および保存

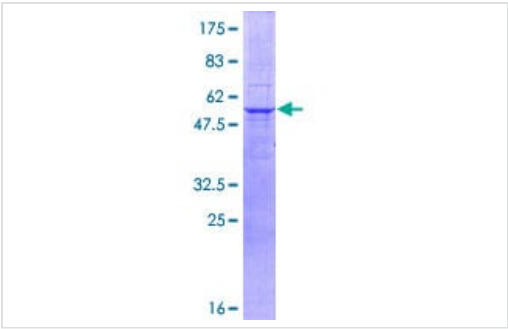
保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

関連情報

機能	Creation of an extracellular membrane face which guides the wrapping process and ultimately compacts adjacent lamellae.
組織特異性	Found only in peripheral nervous system Schwann cells.
関連疾患	Defects in MPZ are the cause of Charcot-Marie-Tooth disease type 1B (CMT1B) [MIM:118200]. CMT1B is a form of Charcot-Marie-Tooth disease, the most common inherited disorder of the peripheral nervous system. Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathy or CMT1, and primary peripheral axonal neuropathy or CMT2. Neuropathies of the CMT1 group are characterized by severely reduced nerve conduction velocities (less than 38 m/sec), segmental demyelination and remyelination with onion bulb formations on nerve biopsy, slowly progressive distal muscle atrophy and weakness, absent deep tendon reflexes, and hollow feet. Defects in MPZ are the cause of Charcot-Marie-Tooth disease type 2I (CMT2I) [MIM:607677]. CMT2I is a form of Charcot-Marie-Tooth disease, the most common inherited disorder of the peripheral nervous system. Charcot-Marie-Tooth disease is classified in two main groups on the basis of electrophysiologic properties and histopathology: primary peripheral demyelinating neuropathy or CMT1, and primary peripheral axonal neuropathy or CMT2. Neuropathies of the CMT2 group are characterized by signs of axonal regeneration in the absence of obvious myelin alterations, normal or slightly reduced nerve conduction velocities, and progressive distal muscle weakness and atrophy. CMT2I is characterized by late onset (range 47 to 60 years). Defects in MPZ are the cause of Charcot-Marie-Tooth disease type 2J (CMT2J) [MIM:607736]. CMT2J is a form of Charcot-Marie-Tooth disease characterized by the association of axonal peripheral neuropathy with hearing loss and pupillary abnormalities such as Adie pupil. Inheritance is autosomal dominant. Defects in MPZ are the cause of Adie pupil (ADIEP) [MIM:103100]. A stationary, benign disorder characterized by tonic, sluggishly reacting pupil and hypoactive or absent tendon reflexes. Adie pupil is a characteristic of Charcot-Marie-Tooth disease type 2J. Defects in MPZ may be the cause of Charcot-Marie-Tooth disease dominant intermediate type D (CMTDID) [MIM:607791]. CMTDID is a form of Charcot-Marie-Tooth disease characterized by features intermediate between demyelinating and axonal peripheral neuropathies, and motor median nerve conduction velocities ranging from 25 to 45 m/sec. Defects in MPZ are a cause of Dejerine-Sottas syndrome (DSS) [MIM:145900]; also known as Dejerine-Sottas neuropathy (DSN) or hereditary motor and sensory neuropathy III (HMSN3). DSS is a severe degenerating neuropathy of the demyelinating Charcot-Marie-Tooth disease category, with onset by age 2 years. DSS is characterized by motor and sensory neuropathy with very slow nerve conduction velocities, increased cerebrospinal fluid protein concentrations, hypertrophic nerve changes, delayed age of walking as well as areflexia. There are both autosomal dominant and autosomal recessive forms of Dejerine-Sottas syndrome. Defects in MPZ are a cause of congenital hypomyelination neuropathy (CHN) [MIM:605253]. CHN is characterized clinically by early onset of hypotonia, areflexia, distal muscle weakness, and very slow nerve conduction velocities. Defects in MPZ are a cause of Roussy-Levy syndrome (ROULS) [MIM:180800]; also known as Roussy-Levy hereditary areflexic dystasia. This autosomal dominant disorder resembles Charcot-

	Marie-Tooth disease type 1 in that it presents with foot deformity, weakness and atrophy of distal limb muscles, especially the peronei, and absent tendon reflexes. The phenotype differs, however, in that it includes static tremor of the upper limbs and gait ataxia.
配列類似性	Belongs to the myelin P0 protein family. Contains 1 Ig-like V-type (immunoglobulin-like) domain.
翻訳後修飾	N-glycosylated; contains sulfate-substituted glycan.
細胞内局在	Membrane.

画像



SDS-PAGE - Recombinant Human Myelin Protein Zero (ab114281)

12.5% SDS-PAGE analysis of Myelin Protein Zero protein (ab114281) Stained with Coomassie Blue.

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