

Title (en)

PROTEIN WHICH INTERACTS WITH THE HUNTINGTON'S DISEASE GENE PRODUCT, cDNA CODING THEREFOR, AND ANTIBODIES THERETO

Title (de)

Bindendes Protein für Genprodukt der Huntingtonschen Krankheit, kodierende cDNA und Antikörper

Title (fr)

PROTEINE AYANT UNE INTERACTION AVEC LE GENE DE LA MALADIE DE HUNTINGTON, ADNc CODANT CETTE PROTEINE ET ANTICORPS CONTRE CELLE-CI

Publication

EP 0873132 A1 19981028 (EN)

Application

EP 96940479 A 19961115

Priority

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Abstract (en)

[origin: WO9718825A1] A protein, designated as HIP1, interacts differently with the gene product of a normal (16 CAG repeat) and an expanded (> 44 CAG repeat) HD gene. The HIP1 protein originally isolated from the yeast two-hybrid screen is encoded by a 1.2 kb cDNA, devoid of stop codons, that is expressed as a 400 amino acid polypeptide. By further screening of a human frontal cortex cDNA library, and employing the protocol for 5' Rapid Amplification of cDNA ends (RACE), a total of 4795 nucleotides (with an open reading frame of 914 amino acids) of the 10 kb message HIP1 have been isolated to date. Expression of the HIP1 protein was found to be limited to the brain, where the interaction of the HIP1 with the HD protein appears to be necessary for the association of the HD protein with the membrane or specific cytoskeletal components to render it functional. Because HIP1 interacts with expanded HD protein less well than with normal length HD, introduction of additional HIP1 or overexpression of HIP-1 can lead to increased functionality of the defective or normal HD protein. Alternatively, modified forms of the HIP1 which bind more effectively to expanded HD could be introduced to convert the expanded HD protein into a functional molecule.

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