Nucleotide sequence of cDNA encoding human cytochrome c oxidase subunit VIc

Mieko Otsuka[1,2], Yoshikuni Mizuno[2], Mitsuo Yoshida[2], Yasuo Kagawa[1] and Shigeo Ohta[1]*

Departments of [1]Biochemistry and [2]Neurology, Jichi Medical School, Minamikawachi-machi, Tochigi-ken 329-04, Japan
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Cytochrome c oxidase is the terminal oxidase in respiratory chain. The defects of this enzyme are often observed in muscle of patients with mitochondrial myopathies. The minor subunits may be defective in some patients as a possibility although the roles of these subunits are unknown. In mammalian cells, the enzyme is composed of 13 different subunits, and the minor subunits of cytochrome c oxidase are encoded by a nuclear genome (1). The cDNA encoding the subunit VIc was cloned from a human fibroblast cDNA library by using a synthetic oligonucleotide, d(ATGAAIGAITTIGAIGAIATG), as a hybridization probe (indicated by a doubled line in the figure). The identical amino acids with the counterparts of beef (2) and rat (3) were underlined. The highly homologous sequence suggests strongly that the cDNA encodes the subunit VIc. It is interesting to note that this mitochondrial protein does not have a presequence which targets mitochondria and that the mature form itself has a property of the presequence, such as some basic amino acid residues distributed in the amino-terminal region (4).

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GGGGGGGGGGGGGTCAGGAAGGACGTTGGTGTTGAGGTTAGCATACGTATCAAGGACAGT  60

AACTACCATGGCTCCCGAAGTTTTGCCAAAACCTCGGATGCGTGGCCTTCTGGCCAGGCG 120
      M  A  P  E  V  L  P  K  P  R  M  R  G  L  L  A  R  R

TCTGCGAAATCATATGGCTGTAGCATTCGTGCTATCCCTGGGGGGTTGCAGCTTTGTATAA 180
 L  R  N  H  M  A  V  A  F  V  L  S  L  G  V  A  A  L  Y  K

GTTTCGTGTGGCTGATCAAAGAAAGAAGGCATACGCAGATTCCTACAGAAACTACGATGT 240
 F  R  V  A  D  Q  R  K  K  A  Y  A  D  S  Y  R  N  Y  D  V

CATGAAAGATTTTGAGGAGATGAGGAAGGCTGGTATCTTTCAGAGTGTAAAGTAATCTTG 300
 M  K  D  F  E  E  M  R  K  A  G  I  F  Q  S  V  K  *

GAATATAAAGAATTTCTTCAGGTTGAATTACCTAGAAGTTTGTCACTGACTTGTGTTCCT 360

GAACTATGCCACATGAATATGTGGGCTAAGAATAGTTCCTCTTGATAAATAAACAATTAA 420

C(A)n
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* To whom correspondence should be addressed.

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