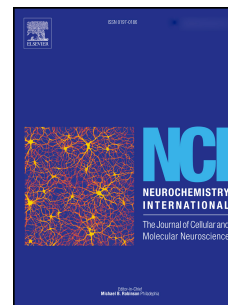


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Brain bioenergetics in rats with acute hyperphenylalaninemia

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α -KGDH, α -ketoglutarate dehydrogenase; ADP, adenosine diphosphate; AMPK, 5' adenosine monophosphate-activated protein kinase; ATP, adenosine triphosphate; BH₄, tetrahydrobiopterin; BSA, bovine serum albumin; CaMKK β , Ca²⁺/calmodulin-dependent kinase kinase β ; CNS, central nervous system; CONCEA, The National Council for Control of Animal Research; Cr, creatine; CS, citrate synthase; CSF, cerebrospinal fluid; ctCK, cytosolic creatine kinase; Drp-1, dynamin-1-like protein; E3, dihydrolipoamide dehydrogenase; EDTA, ethylenediamine tetraacetic acid; EGTA, ethylene glycol tetraacetic acid; GLUT, glucose transporter; HEPES, 4-(2-hydroxyethyl)-1-piperazineethanesulfonic acid; HPA, hyperphenylalaninemia; IDH, isocitrate dehydrogenase; INT, 2(p-iodophenyl)-3-p-nitrophenyl-5-phenyl-tetrazolium chloride; IQ, Intelligence quotient; LAT-1, L-type amino acid transporter 1; LDH, lactate dehydrogenase; MCT, monocarboxylate transporter; MDH, malate dehydrogenase; MFN-1, mitofusin 1; mtCK, mitochondrial creatine kinase; mtDNA, mitochondrial deoxyribonucleic acid; NAD⁺, oxidized nicotinamide adenine dinucleotide; NADH, reduced nicotinamide adenine dinucleotide; NRF-1, nuclear respiratory factor 1; nuDNA, nuclear deoxyribonucleic acid; PAH, phenylalanine hydroxylase; p-Cl-Phe, *p*-chloro-phenylalanine; PCr, phosphocreatine; PDH, pyruvate dehydrogenase; PEP, phosphoenolpyruvate; PGC-1 α , peroxisome proliferator-activated receptor gamma coactivator 1 alpha; Phe, phenylalanine; phospho-AMPK, phosphorylated 5' adenosine monophosphate-activated protein

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