Lack of voltage-dependent anion channel in human mitochondrial myopathies

Str—In several hundreds of patients with a mitochondrial myopathy an enzyme defect in mitochondrial energy metabolism is identified. However, in a substantial number of subjects no enzyme defect can be detected, although diminished substrate oxidation and ATP production rates are found in their muscle in vitro. The hypothesis, that in this group of patients proteins for transport of various ions and substrates across mitochondrial membranes might be affected, led us to study these transport proteins more systematically. Among 40 investigated patients, I was found with a diminished content of the voltage-dependent anion channel (VDAC or human porin).

The patient was born at term from non-consanguineous parents. He had dysmorphism, hypotonia, respiration and feeding problems, and seizures. He was treated for hypothyroidism. Because of lactic acidosis a quadriceps muscle biopsy specimen was taken at the age of 2 years. Decreased rates of pyruvate and malate oxidation and of ATP production were found. The content of the membrane transport proteins VDAC and ATP/ADP translocator (ANT) were estimated immunochemically. The amount of VDAC protein (35 kDa, marked by -v- in figure) appeared to be clearly decreased, with use of monoclonal anti-N-terminus of the membrane-bound ADP/ATP carrier protein 1 Babel D, Walter G, Goetz H, et al. Studies on human porin. VI. Production and characterization of eight monoclonal mouse antibodies against the human VDAC “Porin 31 HL.” and their application for histological studies in human skeletal muscle. Biochim Biophys Acta 1991; 1098: 255-60.

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No aspirin in red wine

Str—Muller and Fugelsang (June 4, p 1428) suggest that wine is a good source of salicylic acid and that this may explain a preventative effect in cardiovascular diseases. We measured salicylic and acetylsalicylic acid in red Bordeaux wines (Rineau 1993, Laveigne 1993, and Mondetour, 1992) with high-performance liquid chromatography and a highly specific fluorescence detection method. The method was validated by varying extraction conditions, mainly extraction solvents and extraction time. In red wine we found 0·7 mg salicylic acid per litre and no acetylsalicylic acid (detection limit 0·025 mg/L). Thus, by contrast with Muller and Fugelsang we found negligible amounts of (acetyl)salicylic acid in wine. Even if wine did contain larger amounts of salicylic acid this would not be expected to affect cardiovascular risk. Salicylic acid and dihydroxybenzoic acids do not affect thromboxane B formation and platelet aggregation; this specifically requires acetylsalicylic acid (aspirin), which inactivates cyclo-oxygenase by irreversible acetylation.

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