

Possible role of apamin-sensitive K⁺ channels in myotonic dystrophy

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Myotonic muscular dystrophy is a genetic disease characterized mainly by muscle atrophy and myotonia, a repetitive electrical activity of muscle. In the present study, the possible role of apamin-sensitive K⁺ channels in the genesis of myotonia was investigated. Apamin is a peptide from bee venom that specifically blocks small conductance Ca²⁺-activated K⁺ channels. The injection of a small amount of apamin (20-30 μ l, 10 μ mol/L) into the thenar muscle of myotonic dystrophy patients decreased the basal electrical activity during the electromyogram in the 6 patients studied. Myotonic discharges after muscle percussion were more difficult to trigger and of smaller intensity and duration. In 2 controls and in 2 patients with generalized myotonia, as well as in 1 patient with myotonia congenita (where the defect is in chloride channels), apamin had no effect. These results suggest that apamin-sensitive K⁺ channels participate in the mechanism that generates myotonia in myotonic dystrophy. ©