

Sarcoglycans in brain

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The sarcoglycan (SG) complex, is composed by six glycosylated transmembrane proteins (α -, β -, δ -, γ -, ϵ - and ζ -SG) and is closely linked to the major dystrophin-glycoprotein complex (DGC). The DGC is essential for membrane integrity during muscle contraction and provides a scaffold for important signalling molecules in all muscle cells. It is well known that Duchenne Muscular Dystrophy is caused by mutations in the dystrophin gene and is characterized by progressive muscle wasting. Cognitive impairments occur in approximately one third of Duchenne patients, highlighting a role for dystrophin in the nervous system. In particular, in neurons, dystrophin and some components of DGC, are associated with inhibitory GABAergic synapses and plays a key role in synapse function and plasticity. In addition, the DGC-complex is involved in blood-brain barrier formation and maintenance and in water and ion homeostasis. Conflicting reports exist concerning possible expression of SGs in brain. Data suggested the presence of ϵ and ζ subunits in CNS; in fact, mutations in the gene encoding ϵ -sarcoglycan cause the neurogenic movement disorder called myoclonus-dystonia, suggesting its role in brain. So the aim of this study is the study of the presence and localization of the SG complex in the human cerebral cortex. Immunofluorescent analysis and reverse transcriptase polymerase chain reaction evaluation were performed for α -, β -, γ -, δ - and ϵ -SG. The results showed that these subunits are expressed in brain. Confocal immunofluorescence microscopy evidenced that all tested proteins of DGC are localized in large neurones in cerebral cortex. An immunoreactivity was also associated with cells that astrocytes. These data suggest that α - β - δ - γ - ϵ -sarcoglycan may have a role in the organization of CNS synapses and in the modulation of blood brain-barrier.