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L345P DES Mutation and Its Influence on the Dynamics of Autophagy Process in Muscle Cells C2C12

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The autophagy process plays an important role in cell cycle, in particular, in the maintenance of proteostasis. In muscle cells, the autophagy process is highly dynamic due to the need for constant updating of the collapsing proteins of the Z-disk area during muscle contraction. In this case, intermediate filaments (IF) play a major role in maintaining the structural and functional integrity of the muscle cell, so the violation of the structure of many of them leads to a change in the dynamics of the autophagy process. In particular, destruction of desmin filaments in C2C12 cell line leads to a significant increase in the rate of degradation of cellular components due to the process of autophagy. It has been shown that in cells with *DES L345P* mutation compared to wild type cells (WT) the amount of LC3-II protein is two times less than in cells with desmin WT after starvation for 2 and 4 h. This increase in the rate of degradation of intracellular components is associated with an increase in the base level of the autophagy process by the removal of mutant forms of desmin protein from the cell, which in the presence of *DES L345P* mutation is polymerizes into aggregates.

Keywords: autophagy, intermediate filaments, muscle cells, desmin, mutations, LC3, *L345P*