

SMNuHomics

Unravelling the Role of SMN in Nucleolar Homeostasis and Potential Implications for Spinal Muscular Atrophy

Spinal Muscular Atrophy (SMA) is a genetic disorder affecting motoneurons, which leads to difficulties in movement, breathing, and swallowing in affected children. SMA is caused by mutations in the SMN1 gene, which encodes a protein called Survival of Motor Neuron (SMN). SMN plays various roles in cellular processes, including the assembly and movement of ribonucleoproteins, which are essential for cell function.

Recent research suggests that SMN also plays a role in the nucleolus, a part of the cell involved in producing components of ribosomes, the cell's protein-making machinery. After DNA repair, SMN moves from its usual location to the nucleolus, where it helps restore its structure. This finding is unexpected and highlights the importance of SMN in maintaining cellular function.

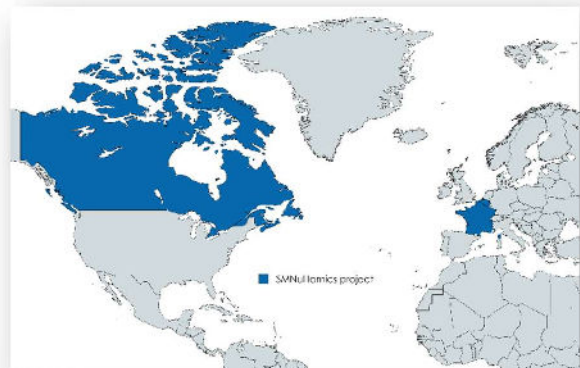
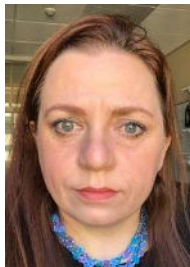
To further understand SMN's role in the nucleolus and its connection to SMA, we have built an international consortium of scientists. We aim to validate SMN's nucleolar function, study how DNA repair affects nucleolar organization in SMA models, and investigate the methylation patterns of proteins interacting with SMN in the nucleolus.

The results of this research could have significant implications for SMA patients, potentially leading to improved diagnostic and prognostic methods and the development of treatments to slow down disease progression and protect motoneurons from degeneration.




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