

Special Issue

Physiological and Pathological Aspects of Unfolded Protein Response

Message from the Guest Editor

Accumulation of unfolded proteins activates the unfolded protein response (UPR), a signalling and transcriptional programme aimed at coping with cellular stress due to misfolding. UPR involves three ER stress sensors, namely PERK, IRE1, and ATF6, which trigger various molecular responses with the aim of reducing protein synthesis and enhancing folding and/or degradation of misfolded proteins. If these measures do not allow the stress to be overcome, the UPR activates apoptotic cell death. Notably, a number of human diseases involve an alteration of proteostasis. The loss of important functions and/or toxic gains of function are the main mechanisms of these misfolding diseases. In the first group of diseases, misshapen proteins never reach their final destination because the ER quality control systems recognize and direct them to disposal (for example, via proteasome or autophagolysosome). In the second group, unfolded proteins aggregate and lead to cellular toxicity through different mechanisms. This overview is not exhaustive but covers possible topics welcome in this Special Issue.

Guest Editor

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