ERp57 as a novel cellular factor controlling prion protein biosynthesis: Therapeutic potential of protein disulfide isomerases

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Resumen

Disturbance of endoplasmic reticulum (ER) proteostasis is observed in Prion-related disorders (PrDs). The protein disulfide isomerase ERp57 is a stress-responsive ER chaperone up-regulated in the brain of Creutzfeldt-Jakob disease patients. However, the actual role of ERp57 in prion protein (PrP) biogenesis and the ER stress response remained poorly defined. We have recently addressed this question using gain- and loss-of-function approaches in vitro and animal models, observing that ERp57 regulates steady-state levels of PrP. Our results revealed that ERp57 modulates the biosynthesis and maturation of PrP but, surprisingly, does not contribute to the global cellular reaction against ER stress in neurons. Here we discuss the relevance of ERp57 as a possible therapeutic target in PrDs and other protein misfolding disorders.

Palabras clave

Palabras clave de autor:PDI; ERp57; ER stress; protein folding; PDIA3; PrP; Grp58
KeyWords Plus:AMYOTROPHIC-LATERAL-SCLEROSIS; ENDOPLASMIC-RETICULUM STRESS; NEURODEGENERATION; REPLICATION;NEUROTOXICITY; PATHOGENESIS; DISEASE

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