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Research background

My research centers around the regulation and function of heat shock proteins (Hsp) in protein quality control. In particular, we study which Hsp can suppress the aggregation and toxicity of disease-associated aggregation inducing proteins. Especially for the proteins with polyglutamine expansion, including mutant huntingtin, we have identified a number of aggregation suppressing Hsp. We study how these Hsp work, how the expression and functional activities of thes HSP are regulated and if and how these can be boosted in possible therapeutic strategies.

Expertise

- HSP function and regulation
- Protein quality control balance (folding, proteasomal degradation, autophagy)
- Protein aggregation and toxicity

Techniques

- cell models for chaperones regulation and protein folding, degradation and autophagy
- biochemical analysis of protein folding and aggregation
- drosophila models of neurodegeneration

Selection of 10 most relevant/recent publications

- Bailey CK Andriola IFM, **Kampinga HH** and Merry DE. Molecular chaperones enhance the degradation of expanded polyglutamine repeat androgen receptor in a cellular model of spinal and bulbar muscular atrophy. **Human Molec. Genet.**, **2002**: 11: 515-523
- Rujano MA, Bosveld A, Salomons FA, Dijk F, van Waarde MAWH, van der Want JJL, de Vos RAI, Brunt ER, Sibon OCM, **Kampinga HH**. Polarized asymmetric inheritance of accumulated protein damage in higher eukaryotes. **PloS Biology**, **2006**, 4, 2325-2335.
- Carra S, Brunsting JF, Lambert H, Landry J, **Kampinga HH**, HSPB8 participates in protein quality control by a non chaperone-like mechanism that requires eIF2alpha phosphorylation. **J Biol Chem**. 284 (**2009**) 5523-5532.
- Hageman J, Rujano MA, van Waarde MAWH, Kakkar V, Dirks D, Govorukhina N, Oosterveld-Hut HJM, Lubsen NH, **Kampinga HH**. A DNAJB Chaperone Subfamily with HDAC-dependent Activities Suppresses Toxic Protein Aggregation**. Molecular Cell** 37 (**2010**) 355-369.
- Zijlstra MP, Rujano MA, van Waarde MAWH, Vis E, Brunt ERP,**Kampinga HH.**, Levels of DNAJB family members (HSP40) correlate with disease onset in patients with spinocerebellar ataxia type 3. Eur. **J. Neuroscience** 32 (**2010**) 760–770.
- Seidel K, den Dunnen WFA, Schultz C, Paulson H, Frank S, de Vos RA, Brunt ERP, Deller T, **Kampinga HH**, Rüb U. Axonal inclusions in spinocerebellar ataxia type 3 (SCA3). **Acta Neuropathologica** 120 (**2010**) 449-460.
- **Kampinga HH**, Craig EA. The HSP70 chaperone machinery: J proteins as drivers of functional specificity. **Nature Reviews Mol. Cell. Biol.** 11 (**2010**) 579-592.
- Vos MJ, Zijlstra MP, Kanon B, van Waarde-Verhagen MAWH, Brunt ERP, Oosterveld-Hut HMJ, Carra S, Sibon OCM, **Kampinga HH**. HSPB7 is the most potent polyQ aggregation suppressor within the HSPB family of molecular chaperones. **Human Molecular Genetics** 19 (**2010**) 4677-4693.
- Carra S, Boncoraglio A, Bart Kanon, Brunsting JF, Minoia M, Rana A, Vos MJ, Seidel K, Sibon OCM, **Kampinga HH**. Identification of the Drosophila ortholog of HSPB8: implication of HSPB8 loss of function in protein folding diseases. **J Biol Chem.** 285 (**2010**) 37811-37822.

- Vos MJ, Zijlstra MP, Carra S, Sibon OCM, **Kampinga HH**. Small heat shock proteins, protein degradation and protein aggregation diseases. **Autophagy**, 7 (**2011**)101-103.