

5. Literaturverzeichnis

- Aitken, A. 1996. 14-3-3 and its possible role in co-ordinating multiple signalling pathways. *Trends Cell Biol.* 6:341-347.
- Andrade, M.A., and P. Bork. 1995. HEAT repeats in the Huntington's disease protein. *Nat Genet.* 11:115-6.
- Barron, L.H., J.P. Warner, M. Porteous, S. Holloway, S. Simpson, R. Davidson, and D.J. Brock. 1993. A study of the Huntington's disease associated trinucleotide repeat in the Scottish population. *J Med Genet.* 30:1003-7.
- Beal, M.F. 2000. Energetics in the pathogenesis of neurodegenerative diseases. *Trends Neurosci.* 23:298-304.
- Bertaux, F., A.H. Sharp, C.A. Ross, H. Lehrach, G.P. Bates, and E. Wanker. 1998. HAP1-huntingtin interactions do not contribute to the molecular pathology in Huntington's disease transgenic mice. *FEBS Lett.* 426:229-32.
- Block-Galarza, J., K.O. Chase, E. Sapp, K.T. Vaughn, R.B. Vallee, M. DiFiglia, and N. Aronin. 1997. Fast transport and retrograde movement of huntingtin and HAP 1 in axons. *Neuroreport.* 8:2247-51.
- Bonifacino, J.S., and A.M. Weissman. 1998. Ubiquitin and the control of protein fate in the secretory and endocytic pathways. *Annu Rev Cell Dev Biol.* 14:19-57.
- Brinkmann, U., R.E. Mattes, and P. Buckel. 1989. High-level expression of recombinant genes in *Escherichia coli* is dependent on the availability of the dnaY gene product. *Gene.* 85:109-14.
- Brodin, L., P. Low, and O. Shupliakov. 2000. Sequential steps in clathrin-mediated synaptic vesicle endocytosis. *Curr Opin Neurobiol.* 10:312-20.
- Burkhard, P., S.V. Strelkov, and J. Stetefeld. 2001. Coiled coils: a highly versatile protein folding motif. *Trends Cell Biol.* 11:82-88.
- Büssow, K., D. Cahill, W. Nietfeld, D. Bancroft, E. Scherzinger, H. Lehrach, and G. Walter. 1998. A method for global protein expression and antibody screening on high-density filters of an arrayed cDNA library. *Nucleic Acids Res.* 26:5007-8.
- Chai, Y., S.L. Koppenhafer, S.J. Shoosmith, M.K. Perez, and H.L. Paulson. 1999. Evidence for proteasome involvement in polyglutamine disease: localization to nuclear

- inclusions in SCA3/MJD and suppression of polyglutamine aggregation in vitro. *Hum Mol Genet.* 8:673-82.
- Charles, V., E. Mezey, P.H. Reddy, A. Dehejia, T.A. Young, M.H. Polymeropoulos, M.J. Brownstein, and D.A. Tagle. 2000. Alpha-synuclein immunoreactivity of huntingtin polyglutamine aggregates in striatum and cortex of Huntington's disease patients and transgenic mouse models. *Neurosci Lett.* 289:29-32.
- Chen, H., S. Fre, V.I. Slepnev, M.R. Capua, K. Takei, M.H. Butler, P.P. Di Fiore, and P. De Camilli. 1998. Epsin is an EH-domain-binding protein implicated in clathrin-mediated endocytosis. *Nature.* 394:793-7.
- Chopra, V.S., M. Metzler, D.M. Rasper, A.E. Engqvist-Goldstein, R. Singaraja, L. Gan, K.M. Fichter, K. McCutcheon, D. Drubin, D.W. Nicholson, and M.R. Hayden. 2000. HIP12 is a non-proapoptotic member of a gene family including HIP1, an interacting protein with huntingtin. *Mamm Genome.* 11:1006-15.
- Conway, K.A., J.D. Harper, and P.T. Lansbury. 1998. Accelerated in vitro fibril formation by a mutant alpha-synuclein linked to early-onset Parkinson disease. *Nat Med.* 4:1318-20.
- Cooper, J.K., G. Schilling, M.F. Peters, W.J. Herring, A.H. Sharp, Z. Kaminsky, J. Masone, F.A. Khan, M. Delanoy, D.R. Borchelt, V.L. Dawson, T.M. Dawson, and C.A. Ross. 1998. Truncated N-terminal fragments of huntingtin with expanded glutamine repeats form nuclear and cytoplasmic aggregates in cell culture. *Hum Mol Genet.* 7:783-90.
- Cummings, C.J., M.A. Mancini, B. Antalffy, D.B. DeFranco, H.T. Orr, and H.Y. Zoghbi. 1998. Chaperone suppression of aggregation and altered subcellular proteasome localization imply protein misfolding in SCA1. *Nat Genet.* 19:148-54.
- Davies, S.W., M. Turmaine, B.A. Cozens, M. DiFiglia, A.H. Sharp, C.A. Ross, E. Scherzinger, E.E. Wanker, L. Mangiarini, and G.P. Bates. 1997. Formation of neuronal intranuclear inclusions underlies the neurological dysfunction in mice transgenic for the HD mutation. *Cell.* 90:537-48.
- Desagher, S., and J.C. Martinou. 2000. Mitochondria as the central control point of apoptosis. *Trends Cell Biol.* 10:369-77.
- Dictenberg, J.B., W. Zimmerman, C.A. Sparks, A. Young, C. Vidair, Y. Zheng, W. Carrington, F.S. Fay, and S.J. Doxsey. 1998. Pericentrin and gamma-tubulin form a protein complex and are organized into a novel lattice at the centrosome. *J Cell Biol.* 141:163-74.

- DiFiglia, M., E. Sapp, K. Chase, C. Schwarz, A. Meloni, C. Young, E. Martin, J.P. Vonsattel, R. Carraway, S.A. Reeves, and et al. 1995. Huntingtin is a cytoplasmic protein associated with vesicles in human and rat brain neurons. *Neuron*. 14:1075-81.
- DiFiglia, M., E. Sapp, K.O. Chase, S.W. Davies, G.P. Bates, J.P. Vonsattel, and N. Aronin. 1997. Aggregation of huntingtin in neuronal intranuclear inclusions and dystrophic neurites in brain. *Science*. 277:1990-3.
- Drewes, G., B. Lichtenberg-Kraag, F. Doring, E.M. Mandelkow, J. Biernat, J. Goris, M. Doree, and E. Mandelkow. 1992. Mitogen activated protein (MAP) kinase transforms tau protein into an Alzheimer-like state. *Embo J*. 11:2131-8.
- Dreyling, M.H., J.A. Martinez-Climent, M. Zheng, J. Mao, J.D. Rowley, and S.K. Bohlander. 1996. The t(10;11)(p13;q14) in the U937 cell line results in the fusion of the AF10 gene and CALM, encoding a new member of the AP-3 clathrin assembly protein family. *Proc Natl Acad Sci U S A*. 93:4804-9.
- Engelender, S., A.H. Sharp, V. Colomer, M.K. Tokito, A. Lanahan, P. Worley, E.L. Holzbaur, and C.A. Ross. 1997. Huntingtin-associated protein 1 (HAP1) interacts with the p150Glued subunit of dyactin. *Hum Mol Genet*. 6:2205-12.
- Engqvist-Goldstein, A.E., M.M. Kessels, V.S. Chopra, M.R. Hayden, and D.G. Drubin. 1999. An actin-binding protein of the Sla2/Huntingtin interacting protein 1 family is a novel component of clathrin-coated pits and vesicles. *J Cell Biol*. 147:1503-18.
- Fenteany, G., R.F. Standaert, W.S. Lane, S. Choi, E.J. Corey, and S.L. Schreiber. 1995. Inhibition of proteasome activities and subunit-specific amino-terminal threonine modification by lactacystin. *Science*. 268:726-31.
- Ford, M.G., B.M. Pearse, M.K. Higgins, Y. Vallis, D.J. Owen, A. Gibson, C.R. Hopkins, P.R. Evans, and H.T. McMahon. 2001. Simultaneous Binding of PtdIns(4,5)P2 and Clathrin by AP180 in the Nucleation of Clathrin Lattices on Membranes. *Science*. 291:1051-1055.
- Gaidarov, I., Q. Chen, J.R. Falck, K.K. Reddy, and J.H. Keen. 1996. A functional phosphatidylinositol 3,4,5-trisphosphate/phosphoinositide binding domain in the clathrin adaptor AP-2 alpha subunit. Implications for the endocytic pathway. *J Biol Chem*. 271:20922-9.
- Gaidarov, I., and J.H. Keen. 1999. Phosphoinositide-AP-2 interactions required for targeting to plasma membrane clathrin-coated pits. *J Cell Biol*. 146:755-64.

- Gething, M.J. 1999. Role and regulation of the ER chaperone BiP. *Semin Cell Dev Biol.* 10:465-72.
- Gossen, M., and H. Bujard. 1992. Tight control of gene expression in mammalian cells by tetracycline- responsive promoters. *Proc Natl Acad Sci U S A.* 89:5547-51.
- Graham, F.L., and A.J. van der Eb. 1973. A new technique for the assay of infectivity of human adenovirus 5 DNA. *Virology.* 52:456-67.
- Graveland, G.A., R.S. Williams, and M. DiFiglia. 1985. Evidence for degenerative and regenerative changes in neostriatal spiny neurons in Huntington's disease. *Science.* 227:770-3.
- Gu, J., C.G. Stephenson, and J. Iadarola. 1994. Recombinant proteins attached to a nickel-NTA column: use in affinity purification of antibodies. *BioTechniques.* 17:257-262.
- Gutekunst, C.-A., A.I. Levey, C.J. Heilman, W.L. Whaley, H. Yi, N.R. Nash, H.D. Rees, J.J. Madden, and S.H. Hersch. 1995. Identification and localization of huntingtin in brain and human lymphoblastoid cell lines with anti-fusion protein antibodies. *Proc. Natl. Acad. Sci.* 92:8710-8714.
- Gutekunst, C.A., S.H. Li, H. Yi, R.J. Ferrante, X.J. Li, and S.M. Hersch. 1998. The cellular and subcellular localization of huntingtin-associated protein 1 (HAP1): comparison with huntingtin in rat and human. *J Neurosci.* 18:7674-86.
- Harper, P.S. 1991. Huntington's disease. W.B. Saunders Co, Ltd, London.
- Hartl, F.U. 1996. Molecular chaperones in cellular protein folding. *Nature.* 381:571-9.
- HDCRG. 1993. A novel gene containing a trinucleotide repeat that is unstable on Huntington's disease chromosomes. *Cell.* 72:971-983.
- Heiser, V., E. Scherzinger, A. Boeddrich, E. Nordhoff, R. Lurz, N. Schugardt, H Lehrach, and E.E. Wanker. 2000. Inhibition of huntingtin fibrillogenesis by specific antibodies and small molecules: implications for Huntington's disease therapy. *Proc Natl Acad Sci U S A.* 97:6739-44.
- Hirst, J., and M.S. Robinson. 1998. Clathrin and adaptors. *Biochemica et Biophysica Acta.* 1404:173-193.
- Holtzman, D.A., S. Yang, and D.G. Drubin. 1993. Synthetic-lethal interactions identify two novel genes, SLA1 and SLA2, that control membrane cytoskeleton assembly in *Saccharomyces cerevisiae*. *J Cell Biol.* 122:635-44.

- Itoh, T., S. Koshihara, T. Kigawa, A. Kikuchi, S. Yokoyama, and T. Takenawa. 2001. Role of the ENTH Domain in Phosphatidylinositol-4,5-Bisphosphate Binding and Endocytosis. *Science*. 291:1047-1051.
- Johnston, J.A., C.L. Ward, and R.R. Kopito. 1998. Aggresomes: a cellular response to misfolded proteins. *J Cell Biol.* 143:1883-98.
- Kalchman, M.A., H.B. Koide, K. McCutcheon, R.K. Graham, K. Nichol, K. Nishiyama, P. Kazemi-Esfarjani, F.C. Lynn, C. Wellington, M. Metzler, Y.P. Goldberg, I. Kanazawa, R.D. Gietz, and M.R. Hayden. 1997. HIP1, a human homologue of *S. cerevisiae* Sla2p, interacts with membrane-associated huntingtin in the brain. *Nat Genet.* 16:44-53.
- Kay, B.K., M. Yamabhai, B. Wendland, and S.D. Emr. 1999. Identification of a novel domain shared by putative components of the endocytic and cytoskeletal machinery. *Protein Sci.* 8:435-8.
- Kazantsev, A., E. Preisinger, A. Dranovsky, D. Goldgaber, and D. Housman. 1999. Insoluble detergent-resistant aggregates form between pathological and nonpathological lengths of polyglutamine in mammalian cells. *Proc Natl Acad Sci U S A.* 96:11404-9.
- Kedersha, N.L., M. Gupta, W. Li, I. Miller, and P. Anderson. 1999. RNA-binding proteins TIA-1 and TIAR link the phosphorylation of eIF-2 alpha to the assembly of mammalian stress granules. *J Cell Biol.* 147:1431-42.
- Klement, I.A., P.J. Skinner, M.D. Kaytor, H. Yi, S.M. Hersch, H.B. Clark, H.Y. Zoghbi, and H.T. Orr. 1998. Ataxin-1 nuclear localization and aggregation: role in polyglutamine-induced disease in SCA1 transgenic mice. *Cell.* 95:41-53.
- Laemmli, U.K. 1970. Cleavage of structural proteins during the assembly of the head of bacteriophage T4. *Nature.* 227:680-685.
- Landschulz, W.H., P.F. Johnson, and S.L. McKnight. 1988. The leucine zipper: a hypothetical structure common to a new class of DNA binding proteins. *Science.* 240:1759-64.
- Layfield, R., J. Fergusson, A. Aitken, J. Lowe, M. Landon, and R.J. Mayer. 1996. Neurofibrillary tangles of Alzheimer's disease brains contain 14-3-3 proteins. *Neurosci Lett.* 209:57-60.

- Li, S.H., S.H. Hosseini, C.A. Gutekunst, S.M. Hersch, R.J. Ferrante, and X.J. Li. 1998. A human HAP1 homologue. Cloning, expression, and interaction with huntingtin. *J Biol Chem.* 273:19220-7.
- Li, X.J., S.H. Li, A.H. Sharp, F.C. Nucifora, Jr., G. Schilling, A. Larahan, P. Worley, S.H. Snyder, and C.A. Ross. 1995. A huntingtin-associated protein enriched in brain with implications for pathology. *Nature.* 378:398-402.
- Lindner, R. 1994. Purification of clathrin-coated vesicles from bovine brain, liver, and adrenal gland. Academic press, New York. 525-530 pp.
- Lunkes, A., and J.L. Mandel. 1998. A cellular model that recapitulates major pathogenic steps of Huntington's disease. *Hum Mol Genet.* 7:1355-61.
- Luthi-Carter, R., A. Strand, N.L. Peters, S.M. Solano, Z.R. Hollingsworth, A.S. Menon, A.S. Frey, B.S. Spektor, E.B. Penney, G. Schilling, C.A. Ross, D.R. Borchelt, S.J. Tapscott, A.B. Young, J.H. Cha, and J.M. Olson. 2000. Decreased expression of striatal signaling genes in a mouse model of Huntington's disease. *Hum Mol Genet.* 9:1259-71.
- Martin, E.J., M. Kim, J. Velier, E. Sapp, H.S. Lee, G. Laforet, L. Won, K. Chase, P.G. Bhide, A. Heller, N. Aronin, and M. DiFiglia. 1999. Analysis of Huntingtin-associated protein 1 in mouse brain and immortalized striatal neurons. *J Comp Neurol.* 403:421-30.
- Martindale, D., A. Hackam, A. Wieczorek, L. Ellerby, C. Wellington, K. McCutcheon, R. Singaraja, P. Kazemi-Esfarjani, R. Devon, S.U. Kim, D.E. Bredesen, F. Tufaro, and M.R. Hayden. 1998. Length of huntingtin and its polyglutamine tract influences localization and frequency of intracellular aggregates. *Nat Genet.* 18:150-4.
- Masters, C.L., G. Simms, N.A. Weinman, G. Multhaup, B.L. McDonald, and K. Beyreuther. 1985. Amyloid plaque core protein in Alzheimer disease and Down syndrome. *Proc Natl Acad Sci U S A.* 82:4245-9.
- McMahon, H.T. 1999. Endocytosis: an assembly protein for clathrin cages. *Curr Biol.* 9:R332-5.
- Muchowski, P.J., G. Schaffar, A. Sittler, E.E. Wanker, M.K. Hayer-Hartl, and F.U. Hartl. 2000. Hsp70 and hsp40 chaperones can inhibit self-assembly of polyglutamine proteins into amyloid-like fibrils. *Proc Natl Acad Sci U S A.* 97:7841-6.

- Nonet, M.L., A.M. Holgado, F. Brewer, C.J. Serpe, B.A. Norbeck, J. Holleran, L. Wei, E. Hartweg, E.M. Jorgensen, and A. Alfonso. 1999. UNC-11, a *Caenorhabditis elegans* AP180 homologue, regulates the size and protein composition of synaptic vesicles. *Mol Biol Cell*. 10:2343-60.
- Nordhoff, E., V. Egelhofer, P. Giavalisco, H. Eickhoff, M. Horn, T. Przewieslik, D. Theiss, U. Schneider, H. Lehrach, and J. Gobom. 2001. Large-gel two-dimensional electrophoresis-matrix assisted laser desorption/ionization-time of flight-mass spectrometry: an analytical challenge for studying complex protein mixtures. *Electrophoresis*. 22:2844-55.
- Ona, V.O., M. Li, J.P. Vonsattel, L.J. Andrews, S.Q. Khan, W.M. Chung, A.S. Frey, A.S. Menon, X.J. Li, P.E. Stieg, J. Yuan, J.B. Penney, A.B. Young, J.H. Cha, and R.M. Friedlander. 1999. Inhibition of caspase-1 slows disease progression in a mouse model of Huntington's disease. *Nature*. 399:263-7.
- Ostrerova, N., L. Petrucelli, M. Farrer, N. Mehta, P. Choi, J. Hardy, and B. Wolozin. 1999. alpha-Synuclein shares physical and functional homology with 14-3-3 proteins. *J Neurosci*. 19:5782-91.
- Owen, D.J., Y. Vallis, M.E. Noble, J.B. Hunter, T.R. Dafforn, P.R. Evans, and H.T. McMahon. 1999. A structural explanation for the binding of multiple ligands by the alpha-adaptin appendage domain. *Cell*. 97:805-15.
- Owen, D.J., Y. Vallis, B.M. Pearse, H.T. McMahon, and P.R. Evans. 2000. The structure and function of the beta 2-adaptin appendage domain. *Embo J*. 19:4216-27.
- Paulson, H.L. 1999. Protein fate in neurodegenerative proteinopathies: polyglutamine diseases join the (mis)fold. *Am J Hum Genet*. 64:339-45.
- Perutz, M.F. 1996. Glutamine repeats and inherited neurodegenerative diseases: molecular aspects. *Curr Opin Struct Biol*. 6:848-58.
- Pieczyk, M., S. Wax, A.R. Beck, N. Kedersha, M. Gupta, B. Maritim, S. Chen, C. Gueydan, V. Kruys, M. Streuli, and P. Anderson. 2000. TIA-1 is a translational silencer that selectively regulates the expression of TNF-alpha. *Embo J*. 19:4154-63.
- Pietromonaco, S.F., G.A. Seluja, A. Aitken, and L. Elias. 1996. Association of 14-3-3 proteins with centrosomes. *Blood Cells Mol Dis*. 22:225-37.

- Plempner, R.K., S. Bohmler, J. Bordallo, T. Sommer, and D.H. Wolf. 1997. Mutant analysis links the translocon and BiP to retrograde protein transport for ER degradation. *Nature*. 388:891-5.
- Polymeropoulos, M.H., C. Lavedan, E. Leroy, S.E. Ide, A. Dehejia, A. Dutra, B. Pike, H. Root, J. Rubenstein, R. Boyer, E.S. Stenroos, S. Chandrasekharappa, A. Athanassiadou, T. Papapetropoulos, W.G. Johnson, A.M. Lazzarini, R.C. Duvoisin, G. Di Iorio, L.I. Golbe, and R.L. Nussbaum. 1997. Mutation in the alpha-synuclein gene identified in families with Parkinson's disease. *Science*. 276:2045-7.
- Reed, T.E., J.H. Chandler, E.M. Hughes, and R.T. Davidson. 1958. Huntington's chorea in Michigan: I. demography and genetics. *American Journal of Human Genetics*. 10:201-225.
- Reynolds, J.P. 1963. The use of lead citrate at high pH as an electron-opaque stain in electron microscopy. *Journal of Cell Biology*. 17:208-212.
- Roehm, N.W., G.H. Rodgers, S.M. Hatfield, and A.L. Glasebrook. 1991. An improved colorimetric assay for cell proliferation and viability utilizing the tetrazolium salt XTT. *J Immunol Methods*. 142:257-65.
- Roizin, L., S. Stellar, and J.C. Liu. 1979. Neuronal Nuclear-Cytoplasmic Changes in Huntington's Chorea: Electron Microscope Investigations. *Advances in Neurology*. 23:95-122.
- Saudou, F., S. Finkbeiner, D. Devys, and M.E. Greenberg. 1998. Huntingtin acts in the nucleus to induce apoptosis but death does not correlate with the formation of intranuclear inclusions. *Cell*. 95:55-66.
- Scherzinger, E., R. Lurz, M. Turmaine, L. Mangiarini, B. Hollenbach, R. Hasenbank, G.P. Bates, S.W. Davies, H. Lehrach, and E.E. Wanker. 1997. Huntingtin-encoded polyglutamine expansions form amyloid-like protein aggregates in vitro and in vivo. *Cell*. 90:549-58.
- Scherzinger, E., A. Sittler, K. Schweiger, V. Heiser, R. Lurz, R. Hasenbank, G.P. Bates, H. Lehrach, and E.E. Wanker. 1999. Self-assembly of polyglutamine-containing huntingtin fragments into amyloid-like fibrils: implications for Huntington's disease pathology. *Proc Natl Acad Sci U S A*. 96:4604-9.
- Schmid, S.L. 1997. Clathrin-coated vesicle formation and protein sorting: an integrated process. *Annu. Rev. Biochem.* 66:511-548.

- Sharp, A.H., S.J. Loev, G. Schilling, S.-H. Li, X.-J. Li, J. Bao, M.V. Wagster, J.A. Kotzok, J.P. Steiner, A. Lo, J. Hedreen, S. Sisodia, S.H. Snyder, T.M. Dawson, D.K. Ryugo, and C.A. Ross. 1995. Widespread expression of Huntington's disease gene (IT15) protein product. *Neuron*. 14:1065-1074.
- Shaw, A. 2000. The 14-3-3 proteins. *Curr Biol*. 10:R400.
- Sherrington, R., E.I. Rogaev, Y. Liang, E.A. Rogaeva, G. Levesque, M. Ikeda, H. Chi, C. Lin, G. Li, K. Holman, and et al. 1995. Cloning of a gene bearing missense mutations in early-onset familial Alzheimer's disease. *Nature*. 375:754-60.
- Sittler, A., R. Lurz, G. Lueder, J. Priller, H. Lehrach, M.K. Hayer-Hartl, F.U. Hartl, and E.E. Wanker. 2001. Geldanamycin activates a heat shock response and inhibits huntingtin aggregation in a cell culture model of Huntington's disease. *Hum Mol Genet*. 10:1307-15.
- Sittler, A., S. Walter, N. Wedemeyer, R. Hasenbank, E. Scherzinger, H. Eickhoff, G.P. Bates, H. Lehrach, and E.E. Wanker. 1998. SH3GL3 associates with the Huntingtin exon 1 protein and promotes the formation of polyglu-containing protein aggregates. *Mol Cell*. 2:427-36.
- Souza, J.M., B.I. Giasson, V.M. Lee, and H. Ischiropoulos. 2000. Chaperone-like activity of synucleins. *FEBS Lett*. 474:116-9.
- Stenoien, D.L., C.J. Cummings, H.P. Adams, M.G. Mancini, K. Patel, G.N. DeMartino, M. Marcelli, N.L. Weigel, and M.A. Mancini. 1999. Polyglutamine-expanded androgen receptors form aggregates that sequester heat shock proteins, proteasome components and SRC-1, and are suppressed by the HDJ-2 chaperone. *Hum Mol Genet*. 8:731-41.
- Steven, A.C., B.L. Trus, J.V. Maizel, M. Unser, D.A. Parry, J.S. Wall, J.F. Hainfeld, and F.W. Studier. 1988. Molecular substructure of a viral receptor-recognition protein. The gp17 tail-fiber of bacteriophage T7. *J Mol Biol*. 200:351-65.
- Stine, O.C., S.H. Li, N. Pleasant, M.V. Wagster, J.C. Hedreen, and C.A. Ross. 1995. Expression of the mutant allele of IT-15 (the HD gene) in striatum and cortex of Huntington's disease patients. *Hum Mol Genet*. 4:15-8.
- Trottier, Y., D. Devys, G. Imbert, F. Sandou, I. An, Y. Lutz, C. Weber, Y. Agid, E.C. Hirsch, and J.-L. Mandel. 1995. Cellular localisation of the Huntington's disease protein and discrimination of the normal and mutated forms. *Nature Genetics*. 10:104-110.

- Tzivion, G., Z.J. Luo, and J. Avruch. 2000. Calyculin A induced vimentin phosphorylation sequesters 14-3-3 and displaces other 14-3-3 partners in vivo. *J Biol Chem.* 275:29772-8.
- Vallee, R.B., and M.P. Sheetz. 1996. Targeting of motor proteins. *Science.* 271:1539-44.
- Velier, J., M. Kim, C. Schwarz, T.W. Kim, E. Sapp, K. Chase, N. Aronin, and M. DiFiglia. 1998. Wild-type and mutant huntingtins function in vesicle trafficking in the secretory and endocytic pathways. *Exp Neurol.* 152:34-40.
- Voges, D., P. Zwickl, and W. Baumeister. 1999. The 26S proteasome: a molecular machine designed for controlled proteolysis. *Annu Rev Biochem.* 68:1015-68.
- Wanker, E.E., C. Rovira, E. Scherzinger, R. Hasenbank, S. Walter, D. Tait, J. Colicelli, and H. Lehrach. 1997. HIP-I: a huntingtin interacting protein isolated by the yeast two-hybrid system. *Hum Mol Genet.* 6:487-95.
- Ward, C.L., S. Omura, and R.R. Kopito. 1995. Degradation of CFTR by the ubiquitin-proteasome pathway. *Cell.* 83:121-7.
- Warrick, J.M., H.Y. Chan, G.L. Gray-Board, Y. Chai, H.L. Paulson, and N.M. Bonini. 1999. Suppression of polyglutamine-mediated neurodegeneration in *Drosophila* by the molecular chaperone HSP70. *Nat Genet.* 23:425-8.
- Wesp, A., L. Hicke, J. Palecek, R. Lombardi, T. Aust, A.L. Munn, and H. Riezman. 1997. End4p/Sla2p interacts with actin-associated proteins for endocytosis in *Saccharomyces cerevisiae*. *Mol Biol Cell.* 8:2291-306.
- Wexler, N.S., A.B. Young, R.E. Tanzi, H. Travers, S. Starosta-Rubinstein, J.B. Penney, S.R. Snodgrass, I. Shoulson, F. Gomez, M.A. Ramos Arroyo, and et al. 1987. Homozygotes for Huntington's disease. *Nature.* 326:194-7.
- Wigley, W.C., R.P. Fabunmi, M.G. Lee, C.R. Marino, S. Muallem, G.N. DeMartino, and P.J. Thomas. 1999. Dynamic association of proteasomal machinery with the centrosome. *J Cell Biol.* 145:481-90.
- Wu, S.Y., and C.M. Chiang. 1996. Establishment of stable cell lines expressing potentially toxic proteins by tetracycline-regulated and epitope-tagging methods. *Biotechniques.* 21:718-22, 724-5.
- Xing, H., S. Zhang, C. Weinheimer, A. Kovacs, and A. Muslin. 2000. 14-3-3 proteins block apoptosis and differentially regulate MAPK cascades. *Embo J.* 19:349-58.

- Yamamoto, A., J.J. Lucas, and R. Hen. 2000. Reversal of neuropathology and motor dysfunction in a conditional model of Huntington's disease. *Cell*. 101:57-66.
- Yang, S., M.J. Cope, and D.G. Drubin. 1999. Sla2p is associated with the yeast cortical actin cytoskeleton via redundant localization signals. *Mol Biol Cell*. 10:2265-83.
- Zhou, S., R. Sousa, N.H. Tannery, and E.M. Lafer. 1992. Characterization of a novel synapse-specific protein. II. cDNA cloning and sequence analysis of the F1-20 protein. *J Neurosci*. 12:2144-55.