

7. Literaturverzeichnis

- Aguzzi, A. and Polymenidou, M. (2004) Mammalian prion biology: one century of evolving concepts. *Cell*, **116**, 313-327.
- Albertinazzi, C., Za, L., Paris, S. and de Curtis, I. (2003) ADP-ribosylation factor 6 and a functional PIX/p95-APP1 complex are required for Rac1B-mediated neurite outgrowth. *Mol Biol Cell*, **14**, 1295-1307.
- Alberts, B. (1998) The cell as a collection of protein machines: preparing the next generation of molecular biologists. *Cell*, **92**, 291-294.
- Alper, T., Haig, D.A. and Clarke, M.C. (1966) The exceptionally small size of the scrapie agent. *Biochem Biophys Res Commun*, **22**, 278-284.
- Andrade, M.A. and Bork, P. (1995) HEAT repeats in the Huntington's disease protein [letter]. *Nat Genet*, **11**, 115-116.
- Arrasate, M., Mitra, S., Schweitzer, E.S., Segal, M.R. and Finkbeiner, S. (2004) Inclusion body formation reduces levels of mutant huntingtin and the risk of neuronal death. *Nature*, **431**, 805-810.
- Bader, G.D., Heilbut, A., Andrews, B., Tyers, M., Hughes, T. and Boone, C. (2003) Functional genomics and proteomics: charting a multidimensional map of the yeast cell. *Trends Cell Biol*, **13**, 344-356.
- Bader, G.D. and Hogue, C.W. (2002) Analyzing yeast protein-protein interaction data obtained from different sources. *Nat Biotechnol*, **20**, 991-997.
- Balbirnie, M., Grothe, R. and Eisenberg, D.S. (2001) An amyloid-forming peptide from the yeast prion Sup35 reveals a dehydrated beta-sheet structure for amyloid. *Proc Natl Acad Sci U S A*, **98**, 2375-2380.
- Barabasi, A.L. and Oltvai, Z.N. (2004) Network biology: understanding the cell's functional organization. *Nat Rev Genet*, **5**, 101-113.
- Bartel, P.L., Chien, C., Sternglanz, R., and Fields, S. (1993) Using the two-hybrid system to detect protein-protein interactions. Oxford: Oxford University Press.
- Bates, G., Harper, S.P. and Jones, L. (2002) *Huntington's Disease*. Oxford University Press.
- Bates, G.P., Mangiarini, L., Mahal, A. and Davies, S.W. (1997) Transgenic models of Huntington's disease. *Hum Mol Genet*, **6**, 1633-1637.
- Baxa, U., Speransky, V., Steven, A.C. and Wickner, R.B. (2002) Mechanism of inactivation on prion conversion of the *Saccharomyces cerevisiae* Ure2 protein. *Proc Natl Acad Sci U S A*, **99**, 5253-5260.

- Beal, M.F., Brouillet, E., Jenkins, B.G., Ferrante, R.J., Kowall, N.W., Miller, J.M., Storey, E., Srivastava, R., Rosen, B.R. and Hyman, B.T. (1993) Neurochemical and histologic characterization of striatal excitotoxic lesions produced by the mitochondrial toxin 3-nitropropionic acid. *J Neurosci*, **13**, 4181-4192.
- Beal, M.F. and Ferrante, R.J. (2004) Experimental therapeutics in transgenic mouse models of Huntington's disease. *Nat Rev Neurosci*, **5**, 373-384.
- Bence, N., Sampat, R. and Kopito, R. (2001) Impairment of the ubiquitin-proteasome system by protein aggregation. *Science*, **292**, 1552-1555.
- Bernstein, C., Bernstein, H., Payne, C.M. and Garewal, H. (2002) DNA repair/proapoptotic dual-role proteins in five major DNA repair pathways: fail-safe protection against carcinogenesis. *Mutat Res*, **511**, 145-178.
- Bessen, R.A., Kocisko, D.A., Raymond, G.J., Nandan, S., Lansbury, P.T. and Caughey, B. (1995) Non-genetic propagation of strain-specific properties of scrapie prion protein. *Nature*, **375**, 698-700.
- Block-Galarza, J., Chase, K.O., Sapp, E., Vaughn, K.T., Vallee, R.B., DiFiglia, M. and Aronin, N. (1997) Fast transport and retrograde movement of huntingtin and HAP 1 in axons. *Neuroreport*, **8**, 2247-2251.
- Bloom, L. and Horvitz, H.R. (1997) The *Caenorhabditis elegans* gene *unc-76* and its human homologs define a new gene family involved in axonal outgrowth and fasciculation. *Proc Natl Acad Sci U S A*, **94**, 3414-3419.
- Boeke, J.D., LaCrute, F. and Fink, G.R. (1984) A positive selection for mutants lacking orotidine-5'-phosphate decarboxylase activity in yeast: 5-fluoro-orotic acid resistance. *Mol Gen Genet*, **197**, 345-346.
- Bonneaud, N., Ozier-Kalogeropoulos, O., Li, G.Y., Labouesse, M., Minvielle-Sebastia, L. and Lacroute, F. (1991) A family of low and high copy replicative, integrative and single-stranded *S. cerevisiae/E. coli* shuttle vectors. *Yeast*, **7**, 609-615.
- Borchsenius, A.S., Wegrzyn, R.D., Newnam, G.P., Inge-Vechtomov, S.G. and Chernoff, Y.O. (2001) Yeast prion protein derivative defective in aggregate shearing and production of new 'seeds'. *Embo J*, **20**, 6683-6691.
- Boutell, J.M., Thomas, P., Neal, J.W., Weston, V.J., Duce, J., Harper, P.S. and Jones, A.L. (1999) Aberrant interactions of transcriptional repressor proteins with the Huntington's disease gene product, huntingtin. *Human Molecular Genetics*, **8**, 1647-1655.
- Bouwmeester, T., Bauch, A., Ruffner, H., Angrand, P.O., Bergamini, G., Croughton, K., Cruciat, C., Eberhard, D., Gagneur, J., Ghidelli, S., Hopf, C., Huhse, B., Mangano, R., Michon, A.M., Schirle, M., Schlegl, J., Schwab, M., Stein, M.A., Bauer, A., Casari, G., Drewes, G., Gavin, A.C., Jackson, D.B., Joberty, G., Neubauer, G., Rick, J., Kuster, B. and Superti-Furga, G. (2004) A physical and functional map of the human TNF-alpha/NF-kappa B signal transduction pathway. *Nat Cell Biol*, **6**, 97-105.

- Bradley, M.E., Edskes, H.K., Hong, J.Y., Wickner, R.B. and Liebman, S.W. (2002) Interactions among prions and prion "strains" in yeast. *Proc Natl Acad Sci U S A*, **99 Suppl 4**, 16392-16399.
- Brouillet, E. and Hantraye, P. (1995) Effects of chronic MPTP and 3-nitropropionic acid in nonhuman primates. *Curr Opin Neurol*, **8**, 469-473.
- Bueler, H., Aguzzi, A., Sailer, A., Greiner, R.A., Autenried, P., Aguet, M. and Weissmann, C. (1993) Mice devoid of PrP are resistant to scrapie. *Cell*, **73**, 1339-1347.
- Bueler, H., Fischer, M., Lang, Y., Bluethmann, H., Lipp, H.P., DeArmond, S.J., Prusiner, S.B., Aguet, M. and Weissmann, C. (1992) Normal development and behaviour of mice lacking the neuronal cell-surface PrP protein. *Nature*, **356**, 577-582.
- Busch, A., Engemann, S., Lurz, R., Okazawa, H., Lehrach, H. and Wanker, E.E. (2003) Mutant huntingtin promotes the fibrillogenesis of wild-type huntingtin: a potential mechanism for loss of huntingtin function in Huntington's disease. *J Biol Chem*.
- Bussow, K., Cahill, D., Nietfeld, W., Bancroft, D., Scherzinger, E., Lehrach, H. and Walter, G. (1998) A method for global protein expression and antibody screening on high- density filters of an arrayed cDNA library. *Nucleic Acids Res*, **26**, 5007-5008.
- Carpenter, D.A. and Ip, W. (1996) Neurofilament triplet protein interactions: evidence for the preferred formation of NF-L-containing dimers and a putative function for the end domains. *J Cell Sci*, **109 (Pt 10)**, 2493-2498.
- Castilla, J., Saa, P., Hetz, C. and Soto, C. (2005) *In vitro* generation of infectious scrapie prions. *Cell*, **121**, 195-206.
- Caughey, B.W., Dong, A., Bhat, K.S., Ernst, D., Hayes, S.F. and Caughey, W.S. (1991) Secondary structure analysis of the scrapie-associated protein PrP 27-30 in water by infrared spectroscopy. *Biochemistry*, **30**, 7672-7680.
- Cepeda, C., Ariano, M.A., Calvert, C.R., Flores-Hernandez, J., Chandler, S.H., Leavitt, B.R., Hayden, M.R. and Levine, M.S. (2001) NMDA receptor function in mouse models of Huntington disease. *J Neurosci Res*, **66**, 525-539.
- Cha, J.H., Kosinski, C.M., Kerner, J.A., Alsdorf, S.A., Mangiarini, L., Davies, S.W., Penney, J.B., Bates, G.P. and Young, A.B. (1998) Altered brain neurotransmitter receptors in transgenic mice expressing a portion of an abnormal human huntington disease gene. *Proc Natl Acad Sci U S A*, **95**, 6480-6485.
- Chandler, R.L. (1961) Encephalopathy in mice produced by inoculation with scrapie brain material. *Lancet*, **1**, 1378-1379.
- Chen, S., Berthelie, V., Yang, W. and Wetzel, R. (2001) Polyglutamine aggregation behavior *in vitro* supports a recruitment mechanism of cytotoxicity. *J Mol Biol*, **311**, 173-182.

- Chernoff, Y.O. (2001) Mutation processes at the protein level: is Lamarck back? *Mutat Res*, **488**, 39-64.
- Chernoff, Y.O., Galkin, A.P., Lewitin, E., Chernova, T.A., Newnam, G.P. and Belenkiy, S.M. (2000) Evolutionary conservation of prion-forming abilities of the yeast Sup35 protein. *Mol Microbiol*, **35**, 865-876.
- Chernoff, Y.O., Lindquist, S.L., Ono, B., Inge-Vechtomov, S.G. and Liebman, S.W. (1995) Role of the chaperone protein Hsp104 in propagation of the yeast prion-like factor [*psi*⁺]. *Science*, **268**, 880-884.
- Chesebro, B., Race, R., Wehrly, K., Nishio, J., Bloom, M., Lechner, D., Bergstrom, S., Robbins, K., Mayer, L., Keith, J.M. and et al. (1985) Identification of scrapie prion protein-specific mRNA in scrapie-infected and uninfected brain. *Nature*, **315**, 331-333.
- Claing, A., Perry, S.J., Achiriloaie, M., Walker, J.K., Albanesi, J.P., Lefkowitz, R.J. and Premont, R.T. (2000) Multiple endocytic pathways of G protein-coupled receptors delineated by GIT1 sensitivity. *Proc Natl Acad Sci U S A*, **97**, 1119-1124.
- Cohen, F.E., Pan, K.M., Huang, Z., Baldwin, M., Fletterick, R.J. and Prusiner, S.B. (1994) Structural clues to prion replication. *Science*, **264**, 530-531.
- Colland, F., Jacq, X., Trouplin, V., Mouglin, C., Groizeleau, C., Hamburger, A., Meil, A., Wojcik, J., Legrain, P. and Gauthier, J.M. (2004) Functional proteomics mapping of a human signaling pathway. *Genome Res*, **14**, 1324-1332.
- Collis, S.J., DeWeese, T.L., Jeggo, P.A. and Parker, A.R. (2005) The life and death of DNA-PK. *Oncogene*, **24**, 949-961.
- Cornett, J., Cao, F., Wang, C.E., Ross, C.A., Bates, G.P., Li, S.H. and Li, X.J. (2005) Polyglutamine expansion of huntingtin impairs its nuclear export. *Nat Genet*, **37**, 198-204.
- Coustou, V., Deleu, C., Saupe, S. and Begueret, J. (1997) The protein product of the *het-s* heterokaryon incompatibility gene of the fungus *Podospora anserina* behaves as a prion analog. *Proc Natl Acad Sci U S A*, **94**, 9773-9778.
- Cox, B.S. (1965) PSI, a cytoplasmic suppressor of super-suppressor in yeast. *Heredity*, **20**, 505-521.
- Cox, B.S., Tuite, M.F. and McLaughlin, C.S. (1988) The *psi* factor of yeast: a problem in inheritance. *Yeast*, **4**, 159-178.
- Damke, H., Baba, T., Warnock, D.E. and Schmid, S.L. (1994) Induction of mutant dynamin specifically blocks endocytic coated vesicle formation. *J Cell Biol*, **127**, 915-934.

- David, G., Abbas, N., Stevanin, G., Durr, A., Yvert, G., Cancel, G., Weber, C., Imbert, G., Saudou, F., Antoniou, E., Drabkin, H., Gemmill, R., Giunti, P., Benomar, A., Wood, N., Ruberg, M., Agid, Y., Mandel, J.L. and Brice, A. (1997) Cloning of the SCA7 gene reveals a highly unstable CAG repeat expansion. *Nat Genet*, **17**, 65-70.
- Davies, S.W., Turmaine, M., Cozens, B.A., DiFiglia, M., Sharp, A.H., Ross, C.A., Scherzinger, E., Wanker, E.E., Mangiarini, L. and Bates, G.P. (1997) Formation of neuronal intranuclear inclusions underlies the neurological dysfunction in mice transgenic for the HD mutation. *Cell*, **90**, 537-548.
- de Curtis, I. (2001) Cell migration: GAPs between membrane traffic and the cytoskeleton. *EMBO Rep*, **2**, 277-281.
- Dechend, R., Hirano, F., Lehmann, K., Heissmeyer, V., Ansieau, S., Wulczyn, F.G., Scheidereit, C. and Leutz, A. (1999) The Bcl-3 oncoprotein acts as a bridging factor between NF-kappaB/Rel and nuclear co-regulators. *Oncogene*, **18**, 3316-3323.
- DePace, A.H., Santoso, A., Hillner, P. and Weissman, J.S. (1998) A critical role for amino-terminal glutamine/asparagine repeats in the formation and propagation of a yeast prion. *Cell*, **93**, 1241-1252.
- Derkatch, I.L., Bradley, M.E., Hong, J.Y. and Liebman, S.W. (2001) Prions affect the appearance of other prions: the story of [PIN(+)]. *Cell*, **106**, 171-182.
- Derkatch, I.L., Bradley, M.E., Masse, S.V., Zadorsky, S.P., Polozkov, G.V., Inge-Vechtomov, S.G. and Liebman, S.W. (2000) Dependence and independence of [PSI⁺] and [PIN⁺]: a two-prion system in yeast? *Embo J*, **19**, 1942-1952.
- Derkatch, I.L., Bradley, M.E., Zhou, P., Chernoff, Y.O. and Liebman, S.W. (1997) Genetic and environmental factors affecting the *de novo* appearance of the [PSI⁺] prion in *Saccharomyces cerevisiae*. *Genetics*, **147**, 507-519.
- Derkatch, I.L., Chernoff, Y.O., Kushnirov, V.V., Inge-Vechtomov, S.G. and Liebman, S.W. (1996) Genesis and variability of [PSI] prion factors in *Saccharomyces cerevisiae*. *Genetics*, **144**, 1375-1386.
- Derkatch, I.L., Uptain, S.M., Outeiro, T.F., Krishnan, R., Lindquist, S.L. and Liebman, S.W. (2004) Effects of Q/N-rich, polyQ, and non-polyQ amyloids on the *de novo* formation of the [PSI⁺] prion in yeast and aggregation of Sup35 *in vitro*. *Proc Natl Acad Sci U S A*, **101**, 12934-12939.
- Di Cesare, A., Paris, S., Albertinazzi, C., Dariozzi, S., Andersen, J., Mann, M., Longhi, R. and de Curtis, I. (2000) p95-APP1 links membrane transport to Rac-mediated reorganization of actin. *Nat Cell Biol*, **2**, 521-530.
- DiFiglia, M. (1997) Clinical Genetics, II. Huntington's disease: from the gene to pathophysiology. *Am J Psychiatry*, **154**, 1046.

- Doel, S.M., McCready, S.J., Nierras, C.R. and Cox, B.S. (1994) The dominant PNM2-mutation which eliminates the psi factor of *Saccharomyces cerevisiae* is the result of a missense mutation in the SUP35 gene. *Genetics*, **137**, 659-670.
- Dohmen, R.J. (2004) SUMO protein modification. *Biochim Biophys Acta*, **1695**, 113-131.
- Dunah, A.W., Jeong, H., Griffin, A., Kim, Y.M., Standaert, D.G., Hersch, S.M., Mouradian, M.M., Young, A.B., Tanese, N. and Krainc, D. (2002) Sp1 and TAFII130 transcriptional activity disrupted in early Huntington's disease. *Science*, **296**, 2238-2243.
- Dure, L.S.t., Young, A.B. and Penney, J.B. (1991) Excitatory amino acid binding sites in the caudate nucleus and frontal cortex of Huntington's disease. *Ann Neurol*, **30**, 785-793.
- Duyao, M.P., Auerbach, A.B., Ryan, A., Persichetti, F., Barnes, G.T., McNeil, S.M., Ge, P., Vonsattel, J.P., Gusella, J.F., Joyner, A.L. and Macdonald, M.E. (1995) Inactivation of the Mouse Huntingtons-Disease Gene Homolog *Hdh*. *Science*, **269**, 407-410.
- Eaglestone, S.S., Ruddock, L.W., Cox, B.S. and Tuite, M.F. (2000) Guanidine hydrochloride blocks a critical step in the propagation of the prion-like determinant $[PSI^+]$ of *Saccharomyces cerevisiae*. *Proc Natl Acad Sci U S A*, **97**, 240-244.
- Ecker, D.J., Khan, M.I., Marsh, J., Butt, T.R. and Crooke, S.T. (1987) Chemical synthesis and expression of a cassette adapted ubiquitin gene. *J. Biol. Chem.*, **262**, 3524-3527.
- Edskes, H.K. and Wickner, R.B. (2002) Conservation of a portion of the *S. cerevisiae* Ure2p prion domain that interacts with the full-length protein. *Proc Natl Acad Sci U S A*, **99 Suppl 4**, 16384-16391.
- Engelender, S., Sharp, A.H., Colomer, V., Tokito, M.K., Lanahan, A., Worley, P., Holzbaur, E.L. and Ross, C.A. (1997) Huntingtin-associated protein 1 (HAP1) interacts with the p150Glued subunit of dynactin. *Hum Mol Genet*, **6**, 2205-2212.
- Faber, P.W., Barnes, G.T., Srinidhi, J., Chen, J., Gusella, J.F. and MacDonald, M.E. (1998) Huntingtin interacts with a family of WW domain proteins. *Hum Mol Genet*, **7**, 1463-1474.
- Ferreira, A. and Rapoport, M. (2002) The synapsins: beyond the regulation of neurotransmitter release. *Cell Mol Life Sci*, **59**, 589-595.
- Ferreira, P.C., Ness, F., Edwards, S.R., Cox, B.S. and Tuite, M.F. (2001) The elimination of the yeast $[PSI^+]$ prion by guanidine hydrochloride is the result of Hsp104 inactivation. *Mol Microbiol*, **40**, 1357-1369.
- Fields, S. and Song, O. (1989) A novel genetic system to detect protein-protein interactions. *Nature*, **340**, 245-246.

- Frolova, L., Le Goff, X., Zhouravleva, G., Davydova, E., Philippe, M. and Kisselev, L. (1996) Eukaryotic polypeptide chain release factor eRF3 is an eRF1- and ribosome-dependent guanosine triphosphatase. *Rna*, **2**, 334-341.
- Garcia, E.P., Mehta, S., Blair, L.A., Wells, D.G., Shang, J., Fukushima, T., Fallon, J.R., Garner, C.C. and Marshall, J. (1998) SAP90 binds and clusters kainate receptors causing incomplete desensitization. *Neuron*, **21**, 727-739.
- Gardian, G. and Vecsei, L. (2004) Huntington's disease: pathomechanism and therapeutic perspectives. *J Neural Transm*, **111**, 1485-1494.
- Ge, H., Walhout, A.J. and Vidal, M. (2003) Integrating 'omic' information: a bridge between genomics and systems biology. *Trends Genet*, **19**, 551-560.
- Gervais, F.G., Singaraja, R., Xanthoudakis, S., Gutekunst, C.A., Leavitt, B.R., Metzler, M., Hackam, A.S., Tam, J., Vaillancourt, J.P., Houtzager, V., Rasper, D.M., Roy, S., Hayden, M.R. and Nicholson, D.W. (2002) Hippi and Hip-1 are molecular accomplices that mediate the recruitment and activation of caspase-8 by an novel mechanism that may contribute to neuronal death in Huntington's disease. *Nature Cell Biology*, **4**, 95-105.
- Gibbs, C.J., Jr., Gajdusek, D.C., Asher, D.M., Alpers, M.P., Beck, E., Daniel, P.M. and Matthews, W.B. (1968) Creutzfeldt-Jakob disease (spongiform encephalopathy): transmission to the chimpanzee. *Science*, **161**, 388-389.
- Gill, G. (2004) SUMO and ubiquitin in the nucleus: different functions, similar mechanisms? *Genes Dev*, **18**, 2046-2059.
- Giot, L., Bader, J.S., Brouwer, C., Chaudhuri, A., Kuang, B., Li, Y., Hao, Y.L., Ooi, C.E., Godwin, B., Vitols, E., Vijayadamar, G., Pochart, P., Machineni, H., Welsh, M., Kong, Y., Zerhusen, B., Malcolm, R., Varrone, Z., Collis, A., Minto, M., Burgess, S., McDaniel, L., Stimpson, E., Spriggs, F., Williams, J., Neurath, K., Ioime, N., Agee, M., Voss, E., Furtak, K., Renzulli, R., Aanensen, N., Carroll, S., Bickelhaupt, E., Lazovatsky, Y., DaSilva, A., Zhong, J., Stanyon, C.A., Finley, R.L., Jr., White, K.P., Braverman, M., Jarvie, T., Gold, S., Leach, M., Knight, J., Shimkets, R.A., McKenna, M.P., Chant, J. and Rothberg, J.M. (2003) A protein interaction map of *Drosophila melanogaster*. *Science*, **302**, 1727-1736.
- Glover, J.R., Kowal, A.S., Schirmer, E.C., Patino, M.M., Liu, J.J. and Lindquist, S. (1997) Self-seeded fibers formed by Sup35, the protein determinant of $[PSI^+]$, a heritable prion-like factor of *S. cerevisiae*. *Cell*, **89**, 811-819.
- Goehler, H., Lalowski, M., Stelzl, U., Waelter, S., Stroedicke, M., Worm, U., Droege, A., Lindenberg, K.S., Knoblich, M., Haenig, C., Herbst, M., Suopanki, J., Scherzinger, E., Abraham, C., Bauer, B., Hasenbank, R., Fritzsche, A., Ludewig, A.H., Bussow, K., Coleman, S.H., Gutekunst, C.A., Landwehrmeyer, B.G., Lehrach, H. and Wanker, E.E. (2004) A protein interaction network links GIT1, an enhancer of huntingtin aggregation, to Huntington's disease. *Mol Cell*, **15**, 853-865.

- Graham, F.L. and van der Eb, A.J. (1973) A new technique for the assay of infectivity of human adenovirus 5 DNA. *Virology*, **52**, 456-467.
- Graveland, G.A., Williams, R.S. and DiFiglia, M. (1985) Evidence for degenerative and regenerative changes in neostriatal spiny neurons in Huntington's disease. *Science*, **227**, 770-773.
- Greene, J.G. and Greenamyre, J.T. (1996) Bioenergetics and glutamate excitotoxicity. *Prog Neurobiol*, **48**, 613-634.
- Griffith, J.S. (1967) Self-replication and scrapie. *Nature*, **215**, 1043-1044.
- Grimminger, V., Richter, K., Imhof, A., Buchner, J. and Walter, S. (2004) The prion curing agent guanidinium chloride specifically inhibits ATP hydrolysis by Hsp104. *J Biol Chem*, **279**, 7378-7383.
- Gutekunst, C.A., Levey, A.I., Heilman, C.J., Whaley, W.L., Yi, H., Nash, N.R., Rees, H.D., Madden, J.J. and Hersch, S.M. (1995) Identification and localization of huntingtin in brain and human lymphoblastoid cell lines with anti-fusion protein antibodies. *Proc Natl Acad Sci U S A*, **92**, 8710-8714.
- Gutekunst, C.-A., Li, S.-H., Yi, H., Mulroy, J.S., Kuemmerle, S., Jones, R., Rye, D., Ferrante, R.J., Hersch, S.M. and Li, X.-J. (1999) Nuclear and neuropil aggregates in Huntington's disease: relationship to neuropathology. *J. Neuroscience*, **19**, 2522-2534.
- Guthrie, C. and Fink, G.R. (1991) *Guide to Yeast Genetics and Molecular Biology*. ACADEMIC PRESS, INC., San Diego.
- Hackam, A.S., Yassa, A.S., Singaraja, R., Metzler, M., Gutekunst, C.A., Gan, L., Warby, S., Wellington, C.L., Vaillancourt, J., Chen, N., Gervais, F.G., Raymond, L., Nicholson, D.W. and Hayden, M.R. (2000) Huntingtin interacting protein 1 induces apoptosis via a novel caspase-dependent death effector domain. *J Biol Chem*, **275**, 41299-41308.
- Harjes, P. and Wanker, E.E. (2003) The hunt for huntingtin function: interaction partners tell many different stories. *Trends Biochem Sci*, **28**, 425-433.
- Hattula, K. and Peranen, J. (2000) FIP-2, a coiled-coil protein, links Huntingtin to Rab8 and modulates cellular morphogenesis. *Curr Biol*, **10**, 1603-1606.
- HDCRG. (1993) A novel gene containing a trinucleotide repeat that is unstable on Huntington's disease chromosomes. *Cell*, **72**, 971-983.
- Heiser, V., Scherzinger, E., Boeddrich, A., Nordhoff, E., Lurz, R., Schugardt, N., Lehrach, H. and Wanker, E.E. (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**, 6739-6744.

- Holbert, S., Denghien, I., Kiechle, T., Rosenblatt, A., Wellington, C., Hayden, M.R., Margolis, R.L., Ross, C.A., Dausset, J., Ferrante, R.J. and Neri, C. (2001) The Gln-Ala repeat transcriptional activator CA150 interacts with huntingtin: neuropathologic and genetic evidence for a role in Huntington's disease pathogenesis. *Proc Natl Acad Sci U S A*, **98**, 1811-1816.
- Holmberg, C.I., Staniszewski, K.E., Mensah, K.N., Matouschek, A. and Morimoto, R.I. (2004) Inefficient degradation of truncated polyglutamine proteins by the proteasome. *Embo J*, **23**, 4307-4318.
- Holz, C., Lueking, A., Bovekamp, L., Gutjahr, C., Bolotina, N., Lehrach, H. and Cahill, D.J. (2001) A human cDNA expression library in yeast enriched for open reading frames. *Genome Res*, **11**, 1730-1735.
- Iizuka, M. and Stillman, B. (1999) Histone acetyltransferase HBO1 interacts with the ORC1 subunit of the human initiator protein. *J Biol Chem*, **274**, 23027-23034.
- Ito, A., Lai, C.H., Zhao, X., Saito, S., Hamilton, M.H., Appella, E. and Yao, T.P. (2001a) p300/CBP-mediated p53 acetylation is commonly induced by p53-activating agents and inhibited by MDM2. *Embo J*, **20**, 1331-1340.
- Ito, T., Chiba, T., Ozawa, R., Yoshida, M., Hattori, M. and Sakaki, Y. (2001b) A comprehensive two-hybrid analysis to explore the yeast protein interactome. *Proc Natl Acad Sci U S A*, **98**, 4569-4574.
- Jackson, G.S., Hosszu, L.L., Power, A., Hill, A.F., Kenney, J., Saibil, H., Craven, C.J., Waltho, J.P., Clarke, A.R. and Collinge, J. (1999) Reversible conversion of monomeric human prion protein between native and fibrillogenic conformations. *Science*, **283**, 1935-1937.
- Jarrett, J.T. and Lansbury Jr, P.T. (1993) Seeding "One-dimensional crystallization" of amyloid: a pathogenic mechanism in Alzheimer's disease and scrapie? *Cell*, **73**, 1055-1058.
- Jeong, H., Mason, S.P., Barabasi, A.L. and Oltvai, Z.N. (2001) Lethality and centrality in protein networks. *Nature*, **411**, 41-42.
- Jimenez-Sanchez, G., Childs, B. and Valle, D. (2001) Human disease genes. *Nature*, **409**, 853-855.
- Kalchman, M.A., Koide, H.B., McCutcheon, K., Graham, R.K., Nichol, K., Nishiyama, K., Kazemi-Esfarjani, P., Lynn, F.C., Wellington, C., Metzler, M., Goldberg, Y.P., Kanazawa, I., Gietz, R.D. and Hayden, M.R. (1997) HIP1, a human homologue of *S. cerevisiae* Sla2p, interacts with membrane-associated huntingtin in the brain. *Nat Genet*, **16**, 44-53.
- Kegel, K.B., Meloni, A.R., Yi, Y., Kim, Y.J., Doyle, E., Cuiffo, B.G., Sapp, E., Wang, Y., Qin, Z.H., Chen, J.D., Nevins, J.R., Aronin, N. and DiFiglia, M. (2002) Huntingtin is present in the nucleus, interacts with the transcriptional corepressor C-terminal binding protein, and represses transcription. *J Biol Chem*, **277**, 7466-7476.

- Kim, J.H., Liao, D., Lau, L.F. and Huganir, R.L. (1998) SynGAP: a synaptic RasGAP that associates with the PSD-95/SAP90 protein family. *Neuron*, **20**, 683-691.
- Kim, S., Ko, J., Shin, H., Lee, J.R., Lim, C., Han, J.H., Altrock, W.D., Garner, C.C., Gundelfinger, E.D., Premont, R.T., Kaang, B.K. and Kim, E. (2003) The GIT family of proteins forms multimers and associates with the presynaptic cytomatrix protein Piccolo. *J Biol Chem*, **278**, 6291-6300.
- King, C.Y. and Diaz-Avalos, R. (2004) Protein-only transmission of three yeast prion strains. *Nature*, **428**, 319-323.
- King, C.Y., Tittmann, P., Gross, H., Gebert, R., Aebi, M. and Wuthrich, K. (1997) Prion-inducing domain 2-114 of yeast Sup35 protein transforms *in vitro* into amyloid-like filaments. *Proc Natl Acad Sci U S A*, **94**, 6618-6622.
- Knop, M., Siegers, K., Pereira, G., Zachariae, W., Winsor, B., Nasmyth, K. and Schiebel, E. (1999) Epitope tagging of yeast genes using a PCR-based strategy: more tags and improved practical routines. *Yeast*, **15**, 963-972.
- Ko, J., Kim, S., Valtschanoff, J.G., Shin, H., Lee, J.R., Sheng, M., Premont, R.T., Weinberg, R.J. and Kim, E. (2003) Interaction between liprin-alpha and GIT1 is required for AMPA receptor targeting. *J Neurosci*, **23**, 1667-1677.
- Kocisko, D.A., Come, J.H., Priola, S.A., Chesebro, B., Raymond, G.J., Lansbury, P.T. and Caughey, B. (1994) Cell-free formation of protease-resistant prion protein. *Nature*, **370**, 471-474.
- Krobitsch, S. and Lindquist, S. (2000) Aggregation of huntingtin in yeast varies with the length of the polyglutamine expansion and the expression of chaperone proteins. *Proc Natl Acad Sci U S A*, **97**, 1589-1594.
- Kryndushkin, D.S., Alexandrov, I.M., Ter-Avanesyan, M.D. and Kushnirov, V.V. (2003) Yeast [PSI⁺] prion aggregates are formed by small Sup35 polymers fragmented by Hsp104. *J Biol Chem*, **278**, 49636-49643.
- Kushnirov, V.V., Kochneva-Pervukhova, N.V., Chechenova, M.B., Frolova, N.S. and Ter-Avanesyan, M.D. (2000) Prion properties of the Sup35 protein of yeast *Pichia methanolica*. *Embo J*, **19**, 324-331.
- Kushnirov, V.V., Ter-Avanesyan, M.D., Didichenko, S.A., Smirnov, V.N., Chernoff, Y.O., Derkach, I.L., Novikova, O.N., Inge-Vechtomov, S.G., Neistat, M.A. and Tolstorukov, II. (1990) Divergence and conservation of SUP2 (SUP35) gene of yeast *Pichia pinus* and *Saccharomyces cerevisiae*. *Yeast*, **6**, 461-472.
- Lacroute, F. (1971) Non-Mendelian mutation allowing ureidosuccinic acid uptake in yeast. *J Bacteriol*, **106**, 519-522.
- Laemmli, U.K. (1970) Cleavage of structural proteins during the assembly of the head of bacteriophage T4. *Nature*, **227**, 680-685.

- Legname, G., Baskakov, I.V., Nguyen, H.O., Riesner, D., Cohen, F.E., DeArmond, S.J. and Prusiner, S.B. (2004) Synthetic mammalian prions. *Science*, **305**, 673-676.
- Li, S., Armstrong, C.M., Bertin, N., Ge, H., Milstein, S., Boxem, M., Vidalain, P.O., Han, J.D., Chesneau, A., Hao, T., Goldberg, D.S., Li, N., Martinez, M., Rual, J.F., Lamesch, P., Xu, L., Tewari, M., Wong, S.L., Zhang, L.V., Berriz, G.F., Jacotot, L., Vaglio, P., Reboul, J., Hirozane-Kishikawa, T., Li, Q., Gabel, H.W., Elewa, A., Baumgartner, B., Rose, D.J., Yu, H., Bosak, S., Sequerra, R., Fraser, A., Mango, S.E., Saxton, W.M., Strome, S., Van Den Heuvel, S., Piano, F., Vandenhaute, J., Sardet, C., Gerstein, M., Doucette-Stamm, L., Gunsalus, K.C., Harper, J.W., Cusick, M.E., Roth, F.P., Hill, D.E. and Vidal, M. (2004) A map of the interactome network of the metazoan *C. elegans*. *Science*, **303**, 540-543.
- Li, S.H., Cheng, A.L., Zhou, H., Lam, S., Rao, M., Li, H. and Li, X.J. (2002) Interaction of Huntington disease protein with transcriptional activator Sp1. *Mol Cell Biol*, **22**, 1277-1287.
- Li, S.H. and Li, X.J. (2004) Huntingtin-protein interactions and the pathogenesis of Huntington's disease. *Trends Genet*, **20**, 146-154.
- Liu, J.J., Sondheimer, N. and Lindquist, S.L. (2002) Changes in the middle region of Sup35 profoundly alter the nature of epigenetic inheritance for the yeast prion [PSI⁺]. *Proc Natl Acad Sci U S A*, **99 Suppl 4**, 16446-16453.
- Lunkes, A., Lindenberg, K.S., Ben-Haiem, L., Weber, C., Devys, D., Landwehrmeyer, G.B., Mandel, J.L. and Trottier, Y. (2002) Proteases acting on mutant huntingtin generate cleaved products that differentially build up cytoplasmic and nuclear inclusions. *Mol Cell*, **10**, 259-269.
- Mangiarini, L., Sathasivam, K., Seller, M., Cozens, B., Harper, A., Hetherington, C., Lawton, M., Trottier, Y., Lehrach, H., Davies, S.W. and Bates, G.P. (1996) Exon 1 of the Huntington's disease gene containing a highly expanded CAG repeat is sufficient to cause a progressive neurological phenotype in transgenic mice. *Cell*, **87**, 493-506.
- Matafora, V., Paris, S., Dariozzi, S. and de Curtis, I. (2001) Molecular mechanisms regulating the subcellular localization of p95-APP1 between the endosomal recycling compartment and sites of actin organization at the cell surface. *J Cell Sci*, **114**, 4509-4520.
- Matus, A. (2000) Actin-based plasticity in dendritic spines. *Science*, **290**, 754-758.
- McKinley, M.P., Bolton, D.C. and Prusiner, S.B. (1983) A protease-resistant protein is a structural component of the scrapie prion. *Cell*, **35**, 57-62.
- Melchior, F., Schergaut, M. and Pichler, A. (2003) SUMO: ligases, isopeptidases and nuclear pores. *Trends Biochem Sci*, **28**, 612-618.

- Metzler, M., Legendre-Guillemain, V., Gan, L., Chopra, V., Kwok, A., McPherson, P.S. and Hayden, M.R. (2001) HIP1 functions in clathrin-mediated endocytosis through binding to clathrin and adaptor protein 2. *J Biol Chem*, **276**, 39271-39276.
- Metzler, M., Li, B., Gan, L., Georgiou, J., Gutekunst, C.A., Wang, Y., Torre, E., Devon, R.S., Oh, R., Legendre-Guillemain, V., Rich, M., Alvarez, C., Gertsenstein, M., McPherson, P.S., Nagy, A., Wang, Y.T., Roder, J.C., Raymond, L.A. and Hayden, M.R. (2003) Disruption of the endocytic protein HIP1 results in neurological deficits and decreased AMPA receptor trafficking. *Embo J*, **22**, 3254-3266.
- Michelitsch, M.D. and Weissman, J.S. (2000) A census of glutamine/asparagine-rich regions: implications for their conserved function and the prediction of novel prions. *Proc Natl Acad Sci U S A*, **97**, 11910-11915.
- Milo, R., Shen-Orr, S., Itzkovitz, S., Kashtan, N., Chklovskii, D. and Alon, U. (2002) Network motifs: simple building blocks of complex networks. *Science*, **298**, 824-827.
- Mitsui, K., Nakayama, H., Akagi, T., Nekooki, M., Ohtawa, K., Takio, K., Hashikawa, T. and Nukina, N. (2002) Purification of polyglutamine aggregates and identification of elongation factor-1alpha and heat shock protein 84 as aggregate-interacting proteins. *J Neurosci*, **22**, 9267-9277.
- Modregger, J., DiProspero, N.A., Charles, V., Tagle, D.A. and Plomann, M. (2002) PACSIN 1 interacts with huntingtin and is absent from synaptic varicosities in presymptomatic Huntington's disease brains. *Hum Mol Genet*, **11**, 2547-2558.
- Muchowski, P.J., Schaffar, G., Sittler, A., Wanker, E.E., Hayer-Hartl, M.K. and Hartl, F.U. (2000) Hsp70 and hsp40 chaperones can inhibit self-assembly of polyglutamine proteins into amyloid-like fibrils. *Proc Natl Acad Sci U S A*, **97**, 7841-7846.
- Mulholland, J., Preuss, D., Moon, A., Wong, A., Drubin, D. and Botstein, D. (1994) Ultrastructure of the yeast actin cytoskeleton and its association with the plasma membrane. *J Cell Biol*, **125**, 381-391.
- Nasir, J., Floresco, S.B., O'Kusky, J.R., Diewert, V.M., Richman, J.M., Zeisler, J., Borowski, A., Marth, J.D., Phillips, A.G. and Hayden, M.R. (1995) Targeted disruption of the Huntington's disease gene results in embryonic lethality and behavioral and morphological changes in heterozygotes. *Cell*, **81**, 811-823.
- Ness, F., Ferreira, P., Cox, B.S. and Tuite, M.F. (2002) Guanidine hydrochloride inhibits the generation of prion "seeds" but not prion protein aggregation in yeast. *Mol Cell Biol*, **22**, 5593-5605.
- Neuwald, A.F. and Hirano, T. (2000) HEAT repeats associated with condensins, cohesins, and other complexes involved in chromosome-related functions. *Genome Res*, **10**, 1445-1452.

- Nucifora, F.C., Sasaki, M., Peters, M.F., Huang, H., Cooper, J.K., Yamada, M., Takahashi, H., Tsuji, S., Troncoso, J., Dawson, V.L., Dawson, T.M. and Ross, C.A. (2001) Interference by Huntingtin and atrophin-1 with CBP-mediated transcription leading to cellular toxicity. *Science*, **291**, 2423-2428.
- Oesch, B., Westaway, D., Walchli, M., McKinley, M.P., Kent, S.B., Aebersold, R., Barry, R.A., Tempst, P., Teplow, D.B., Hood, L.E. and et al. (1985) A cellular gene encodes scrapie PrP 27-30 protein. *Cell*, **40**, 735-746.
- Olney, J.W., Adamo, N.J. and Ratner, A. (1971) Monosodium glutamate effects. *Science*, **172**, 294.
- Ordway, J.M., Tallaksen-Greene, S., Gutekunst, C.A., Bernstein, E.M., Cearley, J.A., Wiener, H.W., Dure, L.S.t., Lindsey, R., Hersch, S.M., Jope, R.S., Albin, R.L. and Detloff, P.J. (1997) Ectopically expressed CAG repeats cause intranuclear inclusions and a progressive late onset neurological phenotype in the mouse. *Cell*, **91**, 753-763.
- Osherovich, L.Z. and Weissman, J.S. (2001) Multiple Gln/Asn-rich prion domains confer susceptibility to induction of the yeast [*PSI*⁺] prion. *Cell*, **106**, 183-194.
- Osherovich, L.Z. and Weissman, J.S. (2002) The utility of prions. *Dev Cell*, **2**, 143-151.
- Owen, D.J., Vallis, Y., Pearse, B.M., McMahon, H.T. and Evans, P.R. (2000) The structure and function of the beta 2-adaptin appendage domain. *Embo J*, **19**, 4216-4227.
- Pan, K.M., Baldwin, M., Nguyen, J., Gasset, M., Serban, A., Groth, D., Mehlhorn, I., Huang, Z., Fletterick, R.J., Cohen, F.E. and et al. (1993) Conversion of alpha-helices into beta-sheets features in the formation of the scrapie prion proteins. *Proc Natl Acad Sci U S A*, **90**, 10962-10966.
- Pantaloni, D. and Carlier, M.F. (1993) How profilin promotes actin filament assembly in the presence of thymosin beta 4. *Cell*, **75**, 1007-1014.
- Paris, S., Longhi, R., Santambrogio, P. and de Curtis, I. (2003) Leucine-zipper-mediated homo- and hetero-dimerization of GIT family p95-ARF GTPase-activating protein, PIX-, paxillin-interacting proteins 1 and 2. *Biochem J*, **372**, 391-398.
- Patino, M.M., Liu, J.J., Glover, J.R. and Lindquist, S. (1996) Support for the prion hypothesis for inheritance of a phenotypic trait in yeast. *Science*, **273**, 622-626.
- Paushkin, S.V., Kushnirov, V.V., Smirnov, V.N. and Ter-Avanesyan, M.D. (1996) Propagation of the yeast prion-like [*psi*⁺] determinant is mediated by oligomerization of the SUP35-encoded polypeptide chain release factor. *Embo J*, **15**, 3127-3134.
- Paushkin, S.V., Kushnirov, V.V., Smirnov, V.N. and Ter-Avanesyan, M.D. (1997) *In vitro* propagation of the prion-like state of yeast Sup35 protein. *Science*, **277**, 381-383.

- Penzes, P., Johnson, R.C., Sattler, R., Zhang, X., Haganir, R.L., Kambampati, V., Mains, R.E. and Eipper, B.A. (2001) The neuronal Rho-GEF Kalirin-7 interacts with PDZ domain-containing proteins and regulates dendritic morphogenesis. *Neuron*, **29**, 229-242.
- Perutz, M.F. (1999) Glutamine repeats and neurodegenerative diseases: molecular aspects. *Trends Biochem Sci*, **24**, 58-63.
- Perutz, M.F., Johnson, T., Suzuki, M. and Finch, J.T. (1994) Glutamine repeats as polar zippers: their possible role in inherited neurodegenerative diseases. *Proc Natl Acad Sci U S A*, **91**, 5355-5358.
- Perutz, M.F., Pope, B.J., Owen, D., Wanker, E.E. and Scherzinger, E. (2002) Aggregation of proteins with expanded glutamine and alanine repeats of the glutamine-rich and asparagine-rich domains of Sup35 and of the amyloid beta-peptide of amyloid plaques. *Proc Natl Acad Sci U S A*, **99**, 5596-5600.
- Poirier, M.A., Li, H., Macosko, J., Cai, S., Amzel, M. and Ross, C.A. (2002) Huntingtin spheroids and protofibrils as precursors in polyglutamine fibrilization. *J Biol Chem*, **277**, 41032-41037.
- Premont, R.T., Claing, A., Vitale, N., Freeman, J.L., Pitcher, J.A., Patton, W.A., Moss, J., Vaughan, M. and Lefkowitz, R.J. (1998) beta2-Adrenergic receptor regulation by GIT1, a G protein-coupled receptor kinase-associated ADP ribosylation factor GTPase-activating protein. *Proc Natl Acad Sci U S A*, **95**, 14082-14087.
- Prusiner, S.B. (1982) Novel proteinaceous infectious particles cause scrapie. *Science*, **216**, 136-144.
- Prusiner, S.B. (1991) Molecular biology of prion diseases. *Science*, **252**, 1515-1522.
- Prusiner, S.B., Groth, D., Serban, A., Koehler, R., Foster, D., Torchia, M., Burton, D., Yang, S.L. and DeArmond, S.J. (1993) Ablation of the prion protein (PrP) gene in mice prevents scrapie and facilitates production of anti-PrP antibodies. *Proc Natl Acad Sci U S A*, **90**, 10608-10612.
- Prusiner, S.B., Hadlow, W.J., Eklund, C.M. and Race, R.E. (1977) Sedimentation properties of the scrapie agent. *Proc Natl Acad Sci U S A*, **74**, 4656-4660.
- Prusiner, S.B., McKinley, M.P., Bowman, K.A., Bolton, D.C., Bendheim, P.E., Groth, D.F. and Glenner, G.G. (1983) Scrapie prions aggregate to form amyloid-like birefringent rods. *Cell*, **35**, 349-358.
- Prusiner, S.B., Scott, M.R., DeArmond, S.J. and Cohen, F.E. (1998) Prion protein biology. *Cell*, **93**, 337-348.
- Ramakers, G.J. (2002) Rho proteins, mental retardation and the cellular basis of cognition. *Trends Neurosci*, **25**, 191-199.

- Ravikumar, B., Vacher, C., Berger, Z., Davies, J.E., Luo, S., Oroz, L.G., Scaravilli, F., Easton, D.F., Duden, R., O'Kane, C.J. and Rubinsztein, D.C. (2004) Inhibition of mTOR induces autophagy and reduces toxicity of polyglutamine expansions in fly and mouse models of Huntington disease. *Nat Genet*, **36**, 585-595.
- Reiner, A., Del Mar, N., Meade, C.A., Yang, H., Dragatsis, I., Zeitlin, S. and Goldowitz, D. (2001) Neurons lacking huntingtin differentially colonize brain and survive in chimeric mice. *J Neurosci*, **21**, 7608-7619.
- Resende, C., Parham, S.N., Tinsley, C., Ferreira, P., Duarte, J.A. and Tuite, M.F. (2002) The *Candida albicans* Sup35p protein (CaSup35p): function, prion-like behaviour and an associated polyglutamine length polymorphism. *Microbiology*, **148**, 1049-1060.
- Ridley, A.J., Paterson, H.F., Johnston, C.L., Diekmann, D. and Hall, A. (1992) The small GTP-binding protein rac regulates growth factor-induced membrane ruffling. *Cell*, **70**, 401-410.
- Rigamonti, D., Bauer, J.H., De-Fraja, C., Conti, L., Sipione, S., Sciorati, C., Clementi, E., Hackam, A., Hayden, M.R., Li, Y., Cooper, J.K., Ross, C.A., Govoni, S., Vincenz, C. and Cattaneo, E. (2000) Wild-type huntingtin protects from apoptosis upstream of caspase-3. *J Neurosci*, **20**, 3705-3713.
- Ringstad, N., Nemoto, Y. and Camilli, P.d. (1997) The SH3p4/SH3p8/SH3p13 protein family: Binding partners for synaptojanin and dynamin via a Grb2-like Src homology 3 domain. *PNAS*, **94**, 8569-8574.
- Ripaud, L., Maillet, L. and Cullin, C. (2003) The mechanisms of [URE3] prion elimination demonstrate that large aggregates of Ure2p are dead-end products. *Embo J*, **22**, 5251-5259.
- Robinson, J.S., Klionsky, D.J., Banta, L.M. and Emr, S.D. (1988) Protein sorting in *Saccharomyces cerevisiae*: isolation of mutants defective in the delivery and processing of multiple vacuolar hydrolases. *Mol Cell Biol*, **8**, 4936-4948.
- Robinson, M.S. (1987) 100-kD coated vesicle proteins: molecular heterogeneity and intracellular distribution studied with monoclonal antibodies. *J Cell Biol*, **104**, 887-895.
- Ross, C.A., Poirier, M.A., Wanker, E.E. and Amzel, M. (2003) Polyglutamine fibrillogenesis: the pathway unfolds. *Proc Natl Acad Sci U S A*, **100**, 1-3.
- Rubinsztein, D.C., Leggo, J., Coles, R., Almqvist, E., Biancalana, V., Cassiman, J.-J., Chotai, K., Connarty, M., Crauford, D., Curtis, A., Curtis, D., Davidson, M.J., Differ, A.-M., Dode, C., Dodge, A., Frontali, M., Ranen, N.G., Stine, O.C., Sherr, M., Abbott, M.H., Franz, M.L., Graham, C.A., Harper, P.S., Hedreen, J.C., Jackson, A., Kaplan, J.-C., Losekoot, M., MacMillan, J.C., Morrison, P., Trottier, Y., Novelletto, A., Simpson, S.A., Theilmann, J., Whittaker, J.L., Folstein, S.E., Ross, C.A. and Hayden, M.R. (1996) Phenotypic characterisation of individuals with 30-40 CAG repeats in the Huntington's disease (HD) gene reveals HD cases with 36 repeats and apparently normal elderly individuals with 36-39 repeats. *American Journal of Human Genetics*, **59**, 16-22.

- Sacchettini, J.C. and Kelly, J.W. (2002) Therapeutic strategies for human amyloid diseases. *Nat Rev Drug Discov*, **1**, 267-275.
- Safar, J., Wille, H., Itri, V., Groth, D., Serban, H., Torchia, M., Cohen, F.E. and Prusiner, S.B. (1998) Eight prion strains have PrP(Sc) molecules with different conformations. *Nat Med*, **4**, 1157-1165.
- Sailer, A., Bueler, H., Fischer, M., Aguzzi, A. and Weissmann, C. (1994) No propagation of prions in mice devoid of PrP. *Cell*, **77**, 967-968.
- Salnikova, A.B., Kryndushkin, D.S., Smirnov, V.N., Kushnirov, V.V. and Ter-Avanesyan, M.D. (2005) Nonsense suppression in yeast cells overproducing Sup35 (eRF3) is caused by its non-heritable amyloids. *J Biol Chem*, **280**, 8808-8812.
- Sambrook, J., Fritsch, E. F., Maniatis, T. (1989) *Molecular Cloning: A Laboratory Manual*. Cold Spring Harbor Laboratory Press.
- Sanchez, I., Mahlke, C. and Yuan, J. (2003) Pivotal role of oligomerization in expanded polyglutamine neurodegenerative disorders. *Nature*, **421**, 373-379.
- Sanger, F., Nicklen, S. and Coulson, A.R. (1977) DNA sequencing with chain-terminating inhibitors. *Proc Natl Acad Sci U S A*, **74**, 5463-5467.
- Santoso, A., Chien, P., Osherovich, L.Z. and Weissman, J.S. (2000) Molecular basis of a yeast prion species barrier. *Cell*, **100**, 277-288.
- Sathasivam, K., Amaechi, I., Mangiarini, L. and Bates, G. (1997) Identification of an HD patient with a (CAG)180 repeat expansion and the propagation of highly expanded CAG repeats in lambda phage. *Hum Genet*, **99**, 692-695.
- Saudou, F., Finkbeiner, S., Devys, D. and Greenberg, M.E. (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., Lurz, R., Turmaine, M., Mangiarini, L., Hollenbach, B., Hasenbank, R., Bates, G.P., Davies, S.W., Lehrach, H. and Wanker, E.E. (1997) Huntingtin-encoded polyglutamine expansions form amyloid-like protein aggregates *in vitro* and *in vivo*. *Cell*, **90**, 549-558.
- Scherzinger, E., Sittler, A., Schweiger, K., Heiser, V., Lurz, R., Hasenbank, R., Bates, G.P., Lehrach, H. and Wanker, E.E. (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**, 4604-4609.
- Schmidt, D. and Muller, S. (2002) Members of the PIAS family act as SUMO ligases for c-Jun and p53 and repress p53 activity. *Proc Natl Acad Sci U S A*, **99**, 2872-2877.
- Schwikowski, B., Uetz, P. and Fields, S. (2000) A network of protein-protein interactions in yeast. *Nat Biotechnol*, **18**, 1257-1261.

- Serio, T.R. and Lindquist, S.L. (2000) Protein-only inheritance in yeast: something to get [PSI⁺]-ched about. *Trends Cell Biol*, **10**, 98-105.
- Sharp, A.H., Loev, S.J., Schilling, G., Li, S.-H., Li, X.-J., Bao, J., Wagster, M.V., Kotzuk, J.A., Steiner, J.P., Lo, A., Hedreen, J., Sisodia, S., Snyder, S.H., Dawson, T.M., Ryugo, D.K. and Ross, C.A. (1995) Widespread expression of Huntington's disease gene (IT15) protein product. *Neuron*, **14**, 1065-1074.
- Sheng, M. and Kim, M.J. (2002) Postsynaptic signaling and plasticity mechanisms. *Science*, **298**, 776-780.
- Sheridan, A.M., Force, T., Yoon, H.J., O'Leary, E., Choukroun, G., Taheri, M.R. and Bonventre, J.V. (2001) PLIP, a novel splice variant of Tip60, interacts with group IV cytosolic phospholipase A(2), induces apoptosis, and potentiates prostaglandin production. *Mol Cell Biol*, **21**, 4470-4481.
- Shorter, J. and Lindquist, S. (2004) Hsp104 catalyzes formation and elimination of self-replicating Sup35 prion conformers. *Science*, **304**, 1793-1797.
- Shorter, J. and Lindquist, S. (2005) Prions as adaptive conduits of memory and inheritance. *Nat Rev Genet*, **6**, 435-450.
- Si, K., Giustetto, M., Etkin, A., Hsu, R., Janisiewicz, A.M., Miniaci, M.C., Kim, J.H., Zhu, H. and Kandel, E.R. (2003a) A neuronal isoform of CPEB regulates local protein synthesis and stabilizes synapse-specific long-term facilitation in aplysia. *Cell*, **115**, 893-904.
- Si, K., Lindquist, S. and Kandel, E.R. (2003b) A neuronal isoform of the aplysia CPEB has prion-like properties. *Cell*, **115**, 879-891.
- Sieradzan, K.A., Mechan, A.O., Jones, L., Wanker, E.E., Nukina, N. and Mann, D.M. (1999) Huntington's disease intranuclear inclusions contain truncated, ubiquitinated huntingtin protein. *Exp Neurol*, **156**, 92-99.
- Singaraja, R.R., Hadano, S., Metzler, M., Givan, S., Wellington, C.L., Warby, S., Yanai, A., Gutekunst, C.A., Leavitt, B.R., Yi, H., Fichter, K., Gan, L., McCutcheon, K., Chopra, V., Michel, J., Hersch, S.M., Ikeda, J.E. and Hayden, M.R. (2002) HIP14, a novel ankyrin domain-containing protein, links huntingtin to intracellular trafficking and endocytosis. *Hum Mol Genet*, **11**, 2815-2828.
- Sittler, A., Walter, S., Wedemeyer, N., Hasenbank, R., Scherzinger, E., Eickhoff, H., Bates, G.P., Lehrach, H. and Wanker, E.E. (1998) SH3GL3 associates with the Huntingtin exon 1 protein and promotes the formation of polygluN-containing protein aggregates. *Mol Cell*, **2**, 427-436.
- Sondheimer, N. and Lindquist, S. (2000) Rnq1: an epigenetic modifier of protein function in yeast. *Mol Cell*, **5**, 163-172.

- Steffan, J.S., Agrawal, N., Pallos, J., Rockabrand, E., Trotman, L.C., Slepko, N., Illes, K., Lukacsovich, T., Zhu, Y.Z., Cattaneo, E., Pandolfi, P.P., Thompson, L.M. and Marsh, J.L. (2004) SUMO modification of Huntingtin and Huntington's disease pathology. *Science*, **304**, 100-104.
- Steffan, J.S., Kazantsev, A., Spasic-Boskovic, O., Greenwald, M., Zhu, Y.Z., Gohler, H., Wanker, E.E., Bates, G.P., Housman, D.E. and Thompson, L.M. (2000) The Huntington's disease protein interacts with p53 and CREB-binding protein and represses transcription. *Proc Natl Acad Sci U S A*, **97**, 6763-6768.
- Sugars, K.L. and Rubinsztein, D.C. (2003) Transcriptional abnormalities in Huntington disease. *Trends Genet*, **19**, 233-238.
- Suhr, S.T., Senut, M.C., Whitelegge, J.P., Faull, K.E., Cuizon, D.B. and Gage, F.H. (2001) Identities of sequestered proteins in aggregates from cells with induced polyglutamine expression. *Journal of Cell Biology*, **153**, 283-294.
- Sun, Y., Savanenin, A., Reddy, P.H. and Liu, Y.F. (2001) Polyglutamine-expanded huntingtin promotes sensitization of N-methyl-D-aspartate receptors via post-synaptic density 95. *J Biol Chem*, **276**, 24713-24718.
- Takekawa, M. and Saito, H. (1998) A family of stress-inducible GADD45-like proteins mediate activation of the stress-responsive MTK1/MEKK4 MAPKKK. *Cell*, **95**, 521-530.
- Tanaka, M., Chien, P., Naber, N., Cooke, R. and Weissman, J.S. (2004) Conformational variations in an infectious protein determine prion strain differences. *Nature*, **428**, 323-328.
- Telling, G.C., Parchi, P., DeArmond, S.J., Cortelli, P., Montagna, P., Gabizon, R., Mastrianni, J., Lugaresi, E., Gambetti, P. and Prusiner, S.B. (1996) Evidence for the conformation of the pathologic isoform of the prion protein enciphering and propagating prion diversity. *Science*, **274**, 2079-2082.
- Ter-Avanesyan, M.D., Dagkesamanskaya, A.R., Kushnirov, V.V. and Smirnov, V.N. (1994) The SUP35 omnipotent suppressor gene is involved in the maintenance of the non-Mendelian determinant [*psi*⁺] in the yeast *Saccharomyces cerevisiae*. *Genetics*, **137**, 671-676.
- Ter-Avanesyan, M.D., Kushnirov, V.V., Dagkesamanskaya, A.R., Didichenko, S.A., Chernoff, Y.O., Inge-Vechtomov, S.G. and Smirnov, V.N. (1993) Deletion analysis of the SUP35 gene of the yeast *Saccharomyces cerevisiae* reveals two non-overlapping functional regions in the encoded protein. *Mol Microbiol*, **7**, 683-692.
- Trottier, Y., Lutz, Y., Stevanin, G., Imbert, G., Devys, D., Cancel, G., Saudou, F., Weber, C., David, G., Tora, L. and et al. (1995) Polyglutamine expansion as a pathological epitope in Huntington's disease and four dominant cerebellar ataxias. *Nature*, **378**, 403-406.

- True, H.L., Berlin, I. and Lindquist, S.L. (2004) Epigenetic regulation of translation reveals hidden genetic variation to produce complex traits. *Nature*, **431**, 184-187.
- True, H.L. and Lindquist, S.L. (2000) A yeast prion provides a mechanism for genetic variation and phenotypic diversity. *Nature*, **407**, 477-483.
- Tuite, M.F., Mundy, C.R. and Cox, B.S. (1981) Agents that cause a high frequency of genetic change from $[psi^+]$ to $[psi^-]$ in *Saccharomyces cerevisiae*. *Genetics*, **98**, 691-711.
- Turner, C.E. (2000) Paxillin interactions. *J Cell Sci*, **113 Pt 23**, 4139-4140.
- Turner, C.E., Brown, M.C., Perrotta, J.A., Riedy, M.C., Nikolopoulos, S.N., McDonald, A.R., Bagrodia, S., Thomas, S. and Leventhal, P.S. (1999) Paxillin LD4 motif binds PAK and PIX through a novel 95-kD ankyrin repeat, ARF-GAP protein: A role in cytoskeletal remodeling. *J Cell Biol*, **145**, 851-863.
- Turner, C.E., West, K.A. and Brown, M.C. (2001) Paxillin-ARF GAP signaling and the cytoskeleton. *Curr Opin Cell Biol*, **13**, 593-599.
- Uetz, P. and Hughes, R.E. (2000) Systematic and large-scale two-hybrid screens. *Curr Opin Microbiol*, **3**, 303-308.
- Vanwetswinkel, S., Kriek, J., Andersen, G.R., Guntert, P., Dijk, J., Canters, G.W. and Siegal, G. (2003) Solution structure of the 162 residue C-terminal domain of human elongation factor 1Bgamma. *J Biol Chem*, **278**, 43443-43451.
- Venkatraman, P., Wetzel, R., Tanaka, M., Nukina, N. and Goldberg, A.L. (2004) Eukaryotic proteasomes cannot digest polyglutamine sequences and release them during degradation of polyglutamine-containing proteins. *Mol Cell*, **14**, 95-104.
- Vitale, N., Patton, W.A., Moss, J., Vaughan, M., Lefkowitz, R.J. and Premont, R.T. (2000) GIT proteins, A novel family of phosphatidylinositol 3,4, 5-trisphosphate-stimulated GTPase-activating proteins for ARF6. *J Biol Chem*, **275**, 13901-13906.
- von Mering, C., Krause, R., Snel, B., Cornell, M., Oliver, S.G., Fields, S. and Bork, P. (2002) Comparative assessment of large-scale data sets of protein-protein interactions. *Nature*, **417**, 399-403.
- Vonsattel, J.-P., Meyers, R.H., Stevens, T.J., Ferrante, R.J., Bird, E.D. and Richardson, E.P. (1985) Neuropathological classification of Huntington's disease. *J. Neuropath. Exp. Neurol.*, **44**, 559-577.
- Waelter, S., Boeddrich, A., Lurz, R., Scherzinger, E., Lueder, G., Lehrach, H. and Wanker, E.E. (2001a) Accumulation of mutant huntingtin fragments in aggregate-like inclusion bodies as a result of insufficient protein degradation. *Mol Biol Cell*, **12**, 1393-1407.

- Waelter, S., Scherzinger, E., Hasenbank, R., Nordhoff, E., Lurz, R., Goehler, H., Gauss, C., Sathasivam, K., Bates, G.P., Lehrach, H. and Wanker, E.E. (2001b) The huntingtin interacting protein HIP1 is a clathrin and alpha-adaptin-binding protein involved in receptor-mediated endocytosis. *Hum Mol Genet*, **10**, 1807-1817.
- Wagster, M.V., Hedreen, J.C., Peyser, C.E., Folstein, S.E. and Ross, C.A. (1994) Selective loss of [3H]kainic acid and [3H]AMPA binding in layer VI of frontal cortex in Huntington's disease. *Exp Neurol*, **127**, 70-75.
- Wang, T., Kobayashi, T., Takimoto, R., Denes, A.E., Snyder, E.L., el-Deiry, W.S. and Brachmann, R.K. (2001) hADA3 is required for p53 activity. *Embo J*, **20**, 6404-6413.
- Wanker, E.E. (2000) Protein aggregation and pathogenesis of Huntington's disease: mechanisms and correlations. *Biol Chem*, **381**, 937-942.
- Wanker, E.E., Rovira, C., Scherzinger, E., Hasenbank, R., Walter, S., Tait, D., Colicelli, J. and Lehrach, H. (1997) HIP-I: a huntingtin interacting protein isolated by the yeast two- hybrid system. *Hum Mol Genet*, **6**, 487-495.
- Wanker, E.E., Scherzinger, E., Heiser, V., Sittler, A., Eickhoff, H. and Lehrach, H. (1999) Membrane filter assay for detection of amyloid-like polyglutamine- containing protein aggregates. *Methods Enzymol*, **309**, 375-386.
- Wegrzyn, R.D., Bapat, K., Newnam, G.P., Zink, A.D. and Chernoff, Y.O. (2001) Mechanism of prion loss after Hsp104 inactivation in yeast. *Mol Cell Biol*, **21**, 4656-4669.
- Wellington, C.L., Singaraja, R., Ellerby, L., Savill, J., Roy, S., Leavitt, B., Cattaneo, E., Hackam, A., Sharp, A., Thornberry, N., Nicholson, D.W., Bredesen, D.E. and Hayden, M.R. (2000) Inhibiting caspase cleavage of huntingtin reduces toxicity and aggregate formation in neuronal and nonneuronal cells. *J Biol Chem*.
- White, J.K., Auerbach, W., Duyao, M.P., Vonsattel, J.P., Gusella, J.F., Joyner, A.L. and MacDonald, M.E. (1997) Huntingtin is required for neurogenesis and is not impaired by the Huntington's disease CAG expansion. *Nat Genet*, **17**, 404-410.
- Wickner, R.B. (1994) [URE3] as an altered URE2 protein: evidence for a prion analog in *Saccharomyces cerevisiae*. *Science*, **264**, 566-569.
- Wickner, R.B., Edskes, H.K., Roberts, B.T., Baxa, U., Pierce, M.M., Ross, E.D. and Brachmann, A. (2004) Prions: proteins as genes and infectious entities. *Genes Dev*, **18**, 470-485.
- Wille, H., Michelitsch, M.D., Guenebaut, V., Supattapone, S., Serban, A., Cohen, F.E., Agard, D.A. and Prusiner, S.B. (2002) Structural studies of the scrapie prion protein by electron crystallography. *Proc Natl Acad Sci U S A*, **99**, 3563-3568.

- Witke, W., Podtelejnikov, A.V., Di Nardo, A., Sutherland, J.D., Gurniak, C.B., Dotti, C. and Mann, M. (1998) In mouse brain profilin I and profilin II associate with regulators of the endocytic pathway and actin assembly. *Embo J*, **17**, 967-976.
- Wu, L.C., Wang, Z.W., Tsan, J.T., Spillman, M.A., Phung, A., Xu, X.L., Yang, M.C., Hwang, L.Y., Bowcock, A.M. and Baer, R. (1996) Identification of a RING protein that can interact *in vivo* with the BRCA1 gene product. *Nat Genet*, **14**, 430-440.
- Yang, W., Dunlap, J.R., Andrews, R.B. and Wetzel, R. (2002) Aggregated polyglutamine peptides delivered to nuclei are toxic to mammalian cells. *Hum Mol Genet*, **11**, 2905-2917.
- Zeitlin, S., Liu, J.P., Chapman, D.L., Papaioannou, V.E. and Efstratiadis, A. (1995) Increased apoptosis and early embryonic lethality in mice nullizygous for the Huntington's disease gene homologue. *Nat Genet*, **11**, 155-163.
- Zhang, H., Webb, D.J., Asmussen, H. and Horwitz, A.F. (2003) Synapse formation is regulated by the signaling adaptor GIT1. *J Cell Biol*, **161**, 131-142.
- Zhang, H., Webb, D.J., Asmussen, H., Niu, S. and Horwitz, A.F. (2005) A GIT1/PIX/Rac/PAK signaling module regulates spine morphogenesis and synapse formation through MLC. *J Neurosci*, **25**, 3379-3388.
- Zhao, Z.S., Manser, E., Loo, T.H. and Lim, L. (2000) Coupling of PAK-interacting exchange factor PIX to GIT1 promotes focal complex disassembly. *Mol Cell Biol*, **20**, 6354-6363.
- Zhouravleva, G., Frolova, L., Le Goff, X., Le Guellec, R., Inge-Vechtormov, S., Kisseliev, L. and Philippe, M. (1995) Termination of translation in eukaryotes is governed by two interacting polypeptide chain release factors, eRF1 and eRF3. *Embo J*, **14**, 4065-4072.
- Zuccato, C., Ciammola, A., Rigamonti, D., Leavitt, B.R., Goffredo, D., Conti, L., MacDonald, M.E., Friedlander, R.M., Silani, V., Hayden, M.R., Timmusk, T., Sipione, S. and Cattaneo, E. (2001) Loss of huntingtin-mediated BDNF gene transcription in Huntington's disease. *Science*, **293**, 493-498.
- Zuccato, C., Tartari, M., Crotti, A., Goffredo, D., Valenza, M., Conti, L., Cataudella, T., Leavitt, B.R., Hayden, M.R., Timmusk, T., Rigamonti, D. and Cattaneo, E. (2003) Huntingtin interacts with REST/NRSF to modulate the transcription of NRSE-controlled neuronal genes. *Nat Genet*, **35**, 76-83.