



**Institut de Neuropatologia  
Hospital Universitari de Bellvitge**

**Departament de Patologia i Terapèutica Experimental  
Universitat de Barcelona**

# **Vías de señalización en enfermedades priónicas**

**Agustín Rodríguez Fernández  
2007**

## 8- Referencias



- Adori C, Kovaks GG, Low P y otros (2005). The ubiquitin-proteasome system in Creutzfeldt-Jakob and Alzheimer disease: intracellular redistribution of components correlates with neuronal vulnerability. *Neurobiol Dis*; 19:427-35.
- Agre P, Kozono D (2003). Aquaporin water channels: molecular mechanisms for human diseases. *FEBS Lett* 555:72-78
- Aguzzi, A. (2003). Prions and the immune system: a journey through gut, spleen, and nerves. *Adv Immunol*. 81:123-71. Review.
- Aguzzi A, Montrasio F, Kaeser PS (2001). Prions: health scare and biological challenge. *Nat Rev Mol Cell Biol* 2:118-126.
- Alper T., Cramp W. A., Haig D. A. and Clarke M. C. (1967). Does the agent of scrapie replicate without nucleic acid? *Nature* 214, 764-766.
- Amiry-Moghaddam M, Williamson A, Palomba M, Eid T, de Lanerolle NC, Naglehus EA, Adams ME, Froehner SC, Agre P, Ottersen OP (2003). Delayed K<sup>+</sup> clearance associated with aquaporin-4 mislocalization: phenotypic defects in brains of alpha-syntrophin-null mice. *Proc Natl Acad Sci USA* 100:13615-13620.
- Amiry-Moghaddam M, Xue R, Haug FM, Neely JD, Bhardwaj A, Agre P, Adams ME, Froehner SC, Mori S, Ottersen OP (2004). Alpha-syntrophin deletion removes the perivascular but endothelial pool of aquaporin-4 at the blood-brain barrier and delays the development of brain edema in an experimental model of acute hyponatremia. *FASEB J* 18:542-544.
- Anderson RM, Donnelly CA, Ferguson NM, et al. (1996). Transmission dynamics and epidemiology of EEB in British cattle. *Nature*; 382: 779-88.
- Andreoletti O, Levavasseur E, Uro-Coste E, Tabouret G, Sarradin P, Delisle M-B, et al. (2002). Astrocytes accumulate 4-hydroxynonenal adducts in murine *scrapie* and human Creutzfeldt-Jakob disease. *Neurobiol Dis*; 11:386-93.
- Angulo E, Casadó V, Mallol J, et al. (2003). A1 Adenosine receptors accumulate in neurodegenerative structures in AD and mediate both

- amyloid precursor protein processing and tau phosphorylation and translocation. *Brain Pathol*; 13:440-51.
- Aoki K, Uchihara T, Tsuchiya K, Nakamura A, Ikeda K, Wakayama Y (2003). Enhanced expression of aquaporin 4 in human brain with infarction. *Acta Neuropathol* 106:121-124.
  - Aoki-Yoshimo K, Uchihara T, Duyckaerts C, Nakamura A, Haw JJ, Wakayama Y (2005). Enhanced expression of aquaporin 4 in human brain with inflammatory diseases. *Acta Neuropathol* 110:281-288.
  - Arrigoni E, Rainnie DG, McCarley RW, et al. (2001). Adenosine-mediated presynaptic modulation of glutamatergic transmission in the laterodorsal tegmentum. *J Neurosci*; 21:1076-85.
  - Badaut J, Lasbennes F, Magistretti PJ, Regli L (2002). Aquaporins in brain: distribution, physiology, and pathophysiology. *J Cereb Blood Flow Metab* 22:367-378.
  - Basler, K., Oesch, B., Scott, M. et al. (1986). *Scrapie* and cellular PrP isoforms are encoded by the same chromosomal gene. *Cell*, 46, 417-442
  - Bio KF, Dal Pizzol F, Conte da Frota ML, Walz R, Andrades ME, da Silva EG, et al. (2001). Imbalance of antioxidant defense in mice lacking cellular prion protein. *Free Radic Biol Med*; 30:1137-44.
  - Bordi F, Ugolini A (1999). Group I metabotropic glutamate receptors: implications for brain diseases. *Progr Neurobiol* 59:55-79.
  - Brandner, S., Isenmann, S., Raeber, A. et al. (1996). Normal host protein necessary for *scrapie*-induced neurotoxicity. *Nature*, 379, 339-34.
  - Bratosiewicz-Wasik J, Wasik TJ, Liberski PP. (2004). Molecular approaches to mechanisms of prion diseases. *Folia Neuropathol*; 42: S33-S46.
  - Brecha S., P. Gass, F. Anton, R. Bravo, M. Zimmermann, T. Herdegen T. (1994). Induction of c-Jun and suppression of CREB transcription factors in axotomized neurons of substantia nigra, and covariation with tyrosine-hydroxylase, *Mol. Cell. Neurosci.* 5 431-441.

- Brown DR, Schulz-Schaeffer WJ, Schmidt B, Krtezschar HA. (1997). Prion protein-deficient cells show altered response to oxidative stress due to decreased SOD-1 activity. *Exp Neurol*; 146:104-12.
- Brown DR, Wong BS, Hafiz F, Clive C, Haswell SJ, Jones IM. (1999). Normal prion protein has an activity like that of superoxide dismutase. *Biochem J*; 344(Pt1):1-5.
- Brown P, Gibbs CJ Jr, Rodgers-Johnson P, et al. (1994). Human spongiform encephalopathy: the National Institutes of Health Series of 300 cases of experimentally transmitted disease. *Ann Neurol*; 35: 513-29.
- Brown P, Preece M, Brandel JP, et al. (2000). Iatrogenic Creutzfeldt-Jakob disease at the millennium. *Neurology*; 55: 1075-81.
- Bruce, M.E. (2003). EET strain variation. *Br. Med. Bull.* 66, 99-108. Review.
- Bruce ME, Will RG, Ironside JW, et al. (1997). Transmissions to mice indicate that 'new variant' ECJ is caused by the EEB agent. *Nature*; 389: 498-01.
- Budka H, Aguzzi A, Brown P, Brucher JM, Bugiani O, Gullotta F, Haltia M, Hauw JJ, Ironside JW, Jellinger K, et al. (1995). Neuropathological diagnostic criteria for Creutzfeldt-Jakob disease (ECJ) and other human spongiform encephalopathies (prion diseases). *Brain Pathol.* 5(4):459-66.
- Budka H, Head MW, Ironside JW, Gambetti P, Parchi P, Zeidler M, Tagliavini F (2003). Sporadic Creutzfeldt-Jakob disease. In: *Neurodegeneration: the molecular pathology of dementia and movement disorders* (Dickson D, ed), pp 287-297. Basel: ISN Neuropath Press.
- Bueler, H., Aguzzi, A., Sailer, A. et al. (1993) Mice devoid of PrP are resistant to *scrapie*. *Cell*, 73, 1339-1347.
- Byung Pal Yu (1994). Cellular defenses against damage from reactive oxygen species. *Physiological reviews* vol. 74, No. 1.

- Capellari S, Cardone F, Notari S, et al. (2005). Creutzfeldt-Jakob disease associated with the R208H mutation in the prion protein gene. *Neurology*; 64: 905-07.
- Caughey B, Lansbury PT. (2003). Protofibrils, pores, fibrils, and neurodegeneration: separating the responsible protein aggregates from the innocent bystanders. *Annu Rev Neurosci*. 26: 267-98.
- Chebib M, Graham ARJ (1999). The "ABC" of GABA receptors: a brief review. *Clin Exp. Pharm Physi* 26: 937-940.
- Choi SI, Ju WK, Choi EK, Kim J, Lea HZ, Carp RI, et al. (1998). Mitochondrial dysfunction induced by oxidative stress in the brains of hamsters infected with the 263K *scrapie* agent. *Acta Neuropathol*; 96:279-86.
- Choi YG, Kim JI, Lee HP, Jin JK, Choi EK, Carp RI, et al. (2000). Induction of heme oxygenase-1 in the brain of *scrapie*-infected mice. *Neurosci Lett*; 289:173-6.
- Ciruela F, Escriche M, Burgueno J, et al. (2001). Metabotropic glutamate 1a and adenosine A1 receptors assemble into functionally interacting complexes; 276:18345-51.
- Clinton J, Forsyth C, Royston MC, Roberts GW (1993). Synaptic degeneration is the primary neuropathological feature in prion diseases: a preliminary study. *Neuroreport* 4: 65-68.
- Collee JG, Bradley R. EEB: a decade on—part II. *Lancet* (1997); 349: 715-21.
- Collee JG, Bradley R. EEB: a decade on—part I. *Lancet* (1997); 349: 636-41;
- Collie DA, Summers DM, Sellar RJ, Ironside JW, Cooper S, Zeidler M, Knight R, Will RG (2003). Diagnosing variant Creutzfeldt- Jakob disease with the pulvinar sign: MR imaging findings in 86 neuropathologically-verified cases. *Am J Neuroradiol* 24:1560-1569.
- Collinge J (2001). Prion diseases of human and animals: their causes and molecular basis. *Annu Rev Neurosci* 24:519-550.

- Conn P, Pin JP (1997). Pharmacology and functions of metabotropic glutamate receptors. *Annu Rev Pharmacol Toxicol* 37:205-237.
- Conn P.J. (2003). Physiological roles and therapeutic potential of metabotropic glutamate receptors, *Ann. N. Y. Acad. Sci.* 1003; 12-21.
- Cowburn RF, O'Neill C, Bonkale WL, Ohm TG, Fastbom J (2001). Receptor G-protein signaling in Alzheimer's disease. *Biochem Soc Symp* 67:163-175.
- Cutting GR, Lu L, Zoghbi H, O'Hara BF, Kasch LM, Montrose-Rafizader C, Donovan DM, Shimada S, Antonarakis SE, Guggino W, Uhl GR, Kazazian HH (1991). Cloning of the  $\gamma$ -aminobutyric acid (GABA) rho 1 cDNA: a GABA receptor subunit highly expressed in the retina. *Proc Natl Acad Sci USA* 88: 2673-2677.
- Dalfó E, Albasanz JL, Martín M, Ferrer I (2004). Abnormal metabotropic glutamate receptor expression and signaling in the cerebral cortex in diffuse Lewy body disease is associated with irregular-synuclein/phospholipase C (PLC $\beta_1$ ) interactions. *Brain Pathol* 14:388-398.
- DeMarco ML, Daggett V. (2005). Local environmental effects on the structure of the prion protein. *C R Biol.* Oct-Nov; 328 (10-11): 847-62. Epub Jun 4. Review.
- Deckert J, Abel F, König G, et al. (1998). Loss of human hippocampal adenosine A1 receptors in dementia: evidence for lack of specificity. *Neurosci Lett*; 244:1-4.
- Deibel MA, Ehmann WD, Markesbery WR. (1996). Copper, iron, and zinc imbalances in severely degenerated brain regions in Alzheimer's disease: possible relation to oxidative stress. *J Neurol Sci.* Nov; 143 (1-2): 137-42.
- Dixon AK, Gubitza AK, Sirinathsinghji DJ, et al. (1996). Tissue distribution of adenosine receptor mRNAs in the rat. *Br J Pharmacol*; 118:461-8.



- Dong CJ, Werblin F S (1994). Dopamine modulation of GABAC receptor function in an isolated retinal neuron. *J Neurophysiol* 71: 1258-1260.
- Dunwiddie TV, Hoffer BJ. (1980). Adenine nucleotides and synaptic transmission in the in vitro rat hippocampus. *Br J Pharmacol*; 69:59-68.
- Dunwiddie TV, Masino SA. (2001). The role and regulation of adenosine in the central nervous system. *Annu Rev Neurosci*; 24:31-55.
- Dunwiddie TV, Masino SA, Poelchen W, et al (2000). Altered electrophysiological sensitivity to A1 but not to GABAB agonists in the hippocampal CA1 region in A1 receptor knockout mice. *Soc Neurosci*; 26:816-8.
- Escriche M, Burgueno J, Ciruela F et al. (2003). Ligand-induced caveolae-mediated internalization of A1 adenosine receptors: morphological evidence of endosomal sorting and receptor recycling. *Exp Cell Res*; 285:72-90.
- Ferrer I, Puig B, Blanco R, Martí E (2000). Prion protein deposition and abnormal synaptic protein expression in the cerebellum in Creutzfeldt-Jakob disease. *Neuroscience* 97:715-726.
- Ferrer I, Rivera R, Blanco R, Marty E (1999). Expression of proteins linked to exocytosis and neurotransmission in patients with Creutzfeldt-Jakob disease. *Neurobiol Dis* 6:92-100.
- Ferrer I. (2002). Differential expression of phosphorylated translation initiator factor 2<sub>e</sub> in Alzheimer's disease and Creutzfeldt-Jakob disease. *Neuropathol Appl Neurobiol*; 28:441-51.
- Ferrer I, B. Friguls, E. Dalfo, A.M. Planas (2003). Early modifications in the expression of mitogen-activated protein kinase (MAPK/ERK), stress-activated kinases SAPK/JNK and p38, and their phosphorylated substrates following focal cerebral ischemia, *Acta Neuropathol.* 105 425-437.
- Ferrer I, G. Santpere, B. Puig, (2006). Immediate early genes, inducible transcription factors, and stress kinases in Alzheimer's

- disease. In: R. Pinaud, L.A. Tremere (Eds.), *Immediate early genes in sensory processing, cognitive performance and neurological disorders*, Springer Science + Business Media, New York, pp. 243-260.
- Ferrer I., R. Blanco, M. Carmona, B. Puig, I. Domínguez, F. Viñals (2002). Active, phosphorylation-dependent MAP kinases, MAPK/ERK, SAPK/JNK and p38, and specific transcription factor substrates are differentially expressed following systemic administration of kainic acid to the adult rat, *Acta Neuropathol.* 103; 391-407.
  - Ferrer I (2005). Pathology of brain edema and brain swelling. In: Kalimo H (ed) *Pathology and genetics of cerebrovascular diseases*. ISN Neuropath Press, Basel, pp 32-38.
  - Floyd CL, Rzigalinski BA, Sitterding HA, Willoughby KA, Ellies EF (2004). Antagonism of group I mGluRs and PLC attenuates increases in IP-3 and reduces reactive gliosis in starving-injured astrocytes. *J Neurotraum* 21:205-216.
  - Fowler JC (1989). Adenosine antagonists delay hypoxia-induced depression of neuronal activity in hippocampal brain slice. *Brain Res*; 490:378-4.
  - Fredholm BB. (1997). Adenosine and neuroprotection. *Int Rev Neurobiol*; 40:259-80.
  - Fredholm BB, Chen JF, Masino SA, et al. (2004). Actions of adenosine at its receptor in the CNS: Insights from knockouts and drugs. *Annu Rev Pharmacol Toxicol*; 45:385-412.
  - Fredholm BB, Dunwiddie TV. (1988). How does adenosine inhibit transmitter release? *Trends Pharmacol Sci*; 9:130-4.
  - Fredholm BB, Ijzerman AP, Jacobson KA, et al. (2001). International union of pharmacology. XXV. Nomenclature and classification of adenosine receptors. *Pharmacol Rev*; 53:527-52.
  - Frigeri A, Gropper MA, Turck CW, Verkman AS (1995) Immunolocalization of the mercurial-insensitive water channel and glycerol intrinsic protein in epithelial cell plasma membranes. *Proc Natl Acad Sci USA* 92:4328-4331

- Fukumitsu N, Ishii K, Kimura Y, et al. (2005). Adenosine A1 receptor mapping of the human brain by PET with 8-dicyclopylelmethyl-1-11C-methyl-3-propylxanthine. *J Nucl Med*; 46:32-7.
- Gajdusek DC, Zigas V. (1957). Degenerative disease of the nervous system in New Guinea: the endemic occurrence of "kuru" in the native population. *N Engl J Med*; 257: 974-78.
- Gambetti P, Parchi P, Chen SG, Cortelli P, Lugaresi E, Montagna P (2003). Fatal insomnia: familial and sporadic. In: *Neurodegeneration: the molecular pathology of dementia and movement disorders* (Dickson D, ed), pp 326-332. Basel: ISN Neuropath Press.
- García-Jimenez A, Fastbom J, Ohm TG, CoWBurn RF (2003). Gprotein  $\alpha$ -subunit levels in the hippocampus and entorhinal cortex of brains staged for Alzheimer's disease neurofibrillary and amyloid pathologies. *Neuroreport* 14:1523-1527.
- Gauthier NS, Headrick JP, Matherne GP. (1998). Myocardial function in the working Mouse heart overexpressing cardiac A1 adenosine receptors. *J Mol Cell Cardiol*; 30:187-93.
- Ghetti B, Bugiani O, Tagliavini F, Piccardo P (2003). Gerstmann-Sträussler-Scheinker disease. In: *Neurodegeneration: the molecular pathology of dementia and movement disorders* (Dickson D, ed), pp 318- 325. Basel: ISN Neuropath Press.
- Giese A, Kretzschmar HA (2001). Prion-induced neuronal damage: the mechanisms of neuronal destruction in the subacute spongiform encephalopathies. *Curr Topics Microbiol Immunol* 253:203-217.
- Glover C.P., D.J. Heywood, A.S. Bienemann, U. Deuschle, J.N. Kew, J.B. Uney, (2004). Adenoviral expression of CREB protects neurons from apoptotic and excitotoxic stress, *Neuroreport* 15; 1171-1175.
- Guentchev M, Siedlak SL, Jarius C, Tagliavini F, Castellani RJ, Perry G, et al. (2002). Oxidative damage to nucleic acids in human prion disease. *Neurobiol Dis*; 9:275-81.

- Guentchev M, Voigtlander T, Haberler C, Groschup MH, Budka H. (2000). Evidence of oxidative stress in experimental prion disease. *Neurobiol Dis*; 7:270-3.
- Gunnarson E, Zelenina M, Aperia A (2004). Regulation of brain aquaporins. *Neuroscience* 129:947-955.
- Guillardon F., T. Skutella, E. Uhlmann, F. Holsboer, M. Zimmermann, C. Behl (1996). Activation of c-Fos contributes to  $\beta$ -amyloid peptide-induced neurotoxicity, *Brain Res.* 706; 268-286.
- Hadlow WJ, Kennedy RC, Race RE. (1982). Natural infection of Suffolk sheep with scrapie virus. *J Infect Dis*; 146: 657-64.
- Hainfellner JA, Liberski PP, Guiroy DC, Cervenakova L, Brown P, Gajdusek DC, Budka H. (1997). *Brain Pathol.* 7(1):547-53.
- Hainfellner JA, Wanschitz J, Jellinger K, Liberski PP, Gullotta F, Budka H. (1998). Coexistence of Alzheimer-type neuropathology in Creutzfeldt-Jakob disease. *Acta Neuropathol (Berl)* 96(2):116-22.
- Hamm HE (1998). The many faces of G protein signaling. *J Biol Chem* 273: 669-672.
- Hammond C (2003). Cellular and molecular neurobiology. In: *The metabotropic glutamate receptors*, pp 314-326. Amsterdam: Academic Press.
- Hartwick AT, Lalonde MR, Barnes S, et al. (2004). Adenosine A1-receptor modulation of glutamate-induced calcium influx in rat retinal ganglion cells. *Invest Ophthalmol Vis Sci*; 45:3740-8.
- Herdegen T., V. Waetzig (2001). AP-1 proteins in the adult brain: facts and fictions about effectors of neuroprotection and neurodegeneration, *Oncogene* 20. 2424-2437.
- Hill AF, Desbruslais M, Joiner S, et al. (1997). The same prion strain causes vECJ and EEB. *Nature*; 389: 448-50.
- Hornabrook RW. (1968). Kuru—a subacute cerebellar degeneration: the natural history and clinical features. *Brain*; 91: 53-74.

- Hourrigan JL, Klingsporn AL. (1996). Scrapie: studies on vertical and horizontal transmission. In: Gibbs CJ Jr, ed. Bovine spongiform encephalopathy: the EEB dilemma. New York: Springer: 59-83.
- Hsiao, K., Baker, H., Crow, T. et al. (1989). Linkage of a prion protein missense variant to Gerstmann–Straussler–Scheinker disease. *Nature*, 338 343-345.
- Huillard d'Aignaux JN, Cousens SN, Maccario J, et al. (2002). The incubation period of kuru. *Epidemiology*; 13: 402-08.
- Hur K, Kim J, Choi S, Choi E-K, Carp RI, Kim Y-S (2002). The pathogenic mechanisms of prion diseases. *Mech Ageing Devel* 123: 1637-1647.
- Jaken S (1996). Protein kinase C isozymes and substrates. *Curr Opin Cell Biol* 8:168-173.
- Jean C. Manson and Nadia L. Tuzi (2001). The murine PrP gene (PRNP) and murine and human PrP proteins. Cambridge University Press 11).
- Jeffrey M, Fraser JR, Halliday BWG, Fowler N, Goodsir CM, Brown DA (1995). Early unsuspected neuron and axon terminal loss in *scrapie*-infected mice revealed by morphometry and immunohistochemistry. *Neuropathol Appl Neurobiol* 21: 41-49.
- Jeffrey M, Halliday WG, Bell J, Johnston AR, MacLeod NK, Ingham C, Sayers AR, Brown DA, Fraser JR (2000). Synapse loss associated with abnormal PrP precedes neuronal degeneration in the *scrapie*-infected murine hippocampus. *Neuropathol Appl Neurobiol* 26: 41-54.
- Jialal, I., G. L. Vega and S. M. Grun (1988). Enzyme systems in rat liver and skeletal muscle. Influences of selenium deficiency, chronic training, and acute exercise. *Arch. Biochem. Biophys.* 263: 150-160.
- Johansson B, Halldner L, Dunwiddie TV, et al. (2001). Hyperalgesia, anxiety, and decreased hypoxic neuroprotection in mice lacking the adenosine A1 receptor. *Proc Natl Acad Sci USA*; 98: 9407-12.
- Johnston JB, Silva C, Gonzalez G et al. (2001). Diminished adenosine A1 receptor expression on macrophages in brain and blood of patients with multiple sclerosis. *Ann Neurol*; 49: 650-8.

- Kiening KL, van Landeghem FK, Schreiber S, Thomale UW, von Deimling A, Unterberg AW, Stover JF (2002). Decreased hemispheric aquaporin-4 is linked to evolving brain edema following controlled cortical impact injury in rats. *Neurosci Lett* 324:105-108.
- Kim JI, Choi SI, Kim NH, Jin JK, Choi EK, Carp RI, et al. (2001). Oxidative stress and neurodegeneration in prion diseases. *Ann N Y Acad Sci*; 928:182-6.
- Kimelberg HK (2004). Water homeostasis in the brain: Basic concepts. *Neuroscience* 129:851-860.
- Kocsis JD, Eng DL, Bhisitkul RB. (1984). Adenosine selectively blocks parallel-fiber-mediated synaptic potentials in rat cerebellar cortex. *Proc Natl Acad Sci USA*; 81:6531-4.
- Kovacs GG, Head MW, Bunn T, Laszlo L, Will RG, Ironside JW. (2000). The role of interleukin 6 in interferon-gamma production in thermally injured mice. *J Neurosci* 20(11):1669-75.
- Kretzschmar HA. (1993). Human prion diseases (spongiform encephalopathies). *Arch. Virol.* 7:261-93 (Suppl.).
- Kropp BS, Degner D, Zerr I, Pilz J, Gleiter CH, Otto M, Rütter E, Kretzschmar HA, Wiltfang J, Kornhuber J, Poser S (2000). Creutzfeldt-Jakob disease and oxidative stress. *Acta Neurol Scand* 101: 332-224.
- Laplanche JL, Delasnerie-Laupretre N, Brandel JP, Chatelain J, Beaudry P, Alperovitch A, Launay JM. (1994). Molecular genetics of prion diseases in France. French Research Group on Epidemiology of Human Spongiform Encephalopathies. *Neurology*; 44(12):2347-51.
- Lasley RD, Narayan P, Uittenbogaard A et al. (2000). Activated cardiac adenosine A(1) receptors translocate out of caveolae. *J Biol Chem*; 275:4417-21.
- Lawson Victoria A., Steven J. Collins, Colin L. Masters and Andrew F. Hill (2005). Prion protein glycosylation. *Journal of Neurochemistry*, 2005, 93, 793-801.

- Leadon, S. A., and P. C. H. ,(1976). Activity in selenium deficient rat liver. *Biochem. Biophys. Res. Commun.* 71: 952-958.
- Lee DW, Sohn HO, Lim YG, Lee HM, Kim YS, Carp RI, et al. (1999). Alteration of free radical metabolism in the brain of mice infected with *scrapie* agent. *Free Radic Res*; 30:499-507.
- Lee TS, Eid T, Mane S, Kim JH, Spencer DD, Ottersen OP, de Lanerolle NC (2004). Aquaporin-4 is increased in the sclerotic hippocampus in human temporal lobe epilepsy. *Acta Neuropathol* 108:493-502.
- Lugaresi E, Medori R, Montagna P, Baruzzi A, Cortelli P, et al. (1986). Fatal familial insomnia and dysautonomia with selective degeneration of the thalamic nuclei. *N. Engl. J. Med.* 315:997-1003.
- MacDonald ST, Sutherland K, Ironside JW. (1996). A quantitative analysis of prion protein immunohistochemical staining in Creutzfeldt-Jakob disease using four anti prion protein antibodies. *Neurodegeneration*; 5(1):87-94.
- MacGibbon G.A., P.A. Lawlor, M. Walton, E. Sirimanne, R.L. Faull, B. Synek, E. Mee, B. Connor, M. Dragunow (1997). Expression of Fos, Jun, and Krox family proteins in Alzheimer's disease, *Exp. Neurol.* 147; 316-332;
- Manley GT, Binder DK, Papadopoulos MC, Verkman AS (2004). New insights into water transport and edema in the central nervous system from phenotype analysis of aquaporin-4 null mice. *Neuroscience* 129:983-991.
- Manley GT, Fujimura M, Ma T, Noshita N, Filiz F, Bollen AW, Chan P, Verkman AS (2000). Aquaporin-4 deletion in mice reduces brain edema after acute water intoxication and ischemic stroke. *Nat Med* 6:159-163.
- Manson Jean C. and Tuzi Nadia L. (2001). The murine PrP gene (*Prnp*), and murine and human PrP proteins. Cambridge University Press 11. Expert reviews in molecular medicine.
- Marcus D.L., J.A. Strafacci, D.C. Miller, S. Masia, C.G. Thomas, J. Rosman, S. Hussain, M.L. Freedman (1998). Quantitative neuronal c-

- fos and c-jun expression in Alzheimer's disease, *Neurobiol. Aging* 19; 393-400.
- Mari L. DeMarco, Valerie Daggett (2005). Local environmental effects on the structure of the prion protein. *C. R. Biologies* 328 847-862.
  - Masters CL, Beyreuther K. (1995). Molecular neuropathology of Alzheimer's disease. *Arzneimittelforschung*. Mar;45(3A):410-2. Review.
  - Matherne GP, Linden J, Byford AM, et al. (1997). Transgenic A1 adenosine receptor overexpression increases myocardial resistance to ischemia. *Proc Natl Acad Sci USA*; 94:6541-6.
  - Medori R, Tritschler HJ, LeBlanc LeBlanc A, Villare F, et al. (1992). Fatal familial insomnia, a prion disease with a mutation at codon 178 of the prion protein gene. *N. Engl. J. Med.* 326:444-49.
  - Mellor H, Parker PJ (1998). The extended protein kinase C superfamily. *Biochem J* 332:281-292.
  - Meyer PT, Elmenhorst D, Bier D, et al. (2005). Quantification of cerebral A1 adenosine receptors in human using [18F] CFPX and PET: an equilibrium approach. *Neuroimage*; 24:1192-04.
  - Michelle M. Aarts, Mark Arundine and Michael Tymianski (2003). Interactions of glutamate receptors with postsynaptic density proteins. *Expert Reviews in Molecular Medicine* vol. 5; 16.
  - Michaelis EK (1998). Molecular biology of glutamate receptors in the central nervous system and their role in excitotoxicity, oxidative stress and aging. *Prog Neurobiol* 54:369-415.
  - Milhavet O, Lehmann S. (2002). Oxidative stress and the prion protein in transmissible spongiform encephalopathies. *Brain Res Rev*; 38:328-39.
  - Mitrová E, Belay G (2002). Creutzfeldt-Jakob disease with E200K mutation in Slovakia: characterization and development. *Acta Virol* 46:31-39.



- Morgan, J.I., T. Curran (1991). Stimulus-transcription coupling in the nervous system. Involvement of the inducible proto-oncogenes fos and jun, *Ann. Rev. Neurosci.* 14; 421-451.
- Murata T, Shiga Y, Higano S, Takahashi S, Mugikura S (2002). Conspicuity and evolution of lesions in Creutzfeldt-Jakob disease at diffusion-weighted imaging. *Am J Neuroradiol* 23:1164-1172.
- Neely JD, Christensen BM, Nilesen S, Agre P (1999). Heterotetrameric composition of aquaporin-4 water channels. *Biochemistry* 38:11156-11163.
- Nielsen S, Nagelhus EA, Amiry-Moghaddam M, Bourque C, Agre P, Ottersen OP (1997). Specialized membrane domains for water transport in glial cells: high-resolution immunogold cytochemistry of aquaporin-4 in rat brain. *J Neurosci* 17:171-180.
- Nieto-Bodelón M., G. Santpere, B. Torrejón-Escribano, B. Puig, I. Ferrer (2006). Expression of transcription factors c-Fos, c-Jun, CREB-1 and ATF-2, and caspase-3 in relation with abnormal tau deposits in Pick's disease, *Acta Neuropathol.* 111; 341-350.
- Nishizuka Y (1992). Intracellular signaling by hydrolysis of phospholipids and activation of protein kinase C. *Science* 258:607-614.
- Oesch B, Westaway D, Walchli M, Mckihley MP, Kent SBH, et al. (1985). A cellular gene encodes scrapie PrP 27-30 protein *Cell* 40: 735-46.
- Oliveira Belebóni R, Oliveira Gomes Carolino R, Baldocchi Pizzo A, Castellan-Baldan L, Coutinho-Netto J, Ferreira dos Santos W, Cysne Coimbra N (2004). Pharmacological and biochemical aspects of GABAergic neurotransmission: Pathological and neuropsychobiological relationships. *Cell Mol Neurobiol* 24: 707-728.
- Oshio K, Binder DK, Liang Y, Bollen A, Feuerstein B, Berger MS, Manley GT (2005). Expression of the aquaporin-1 water channel in human glial tumors. *Neurosurgery* 56:375-381.
- Ozawa S, Kamiya H, Tsuzuki K (1998). Glutamate receptors in the mammalian central nervous system. *Prog Neurobiol* 54:518-618.

- Parchi P, Giese A, Capellari S, Brown P, Schulz-Schaeffer W, Windl O, Zerr I, Budka H, Kopp N, Piccardo P, Poser S, Rojiani A, Streichemberger N, Julien J, Vital C, Ghetti B, Gambetti P, Kretzschmar H (1999). Classification of sporadic Creutzfeldt-Jakob disease based on molecular and phenotypic analysis of 300 subjects. *Ann Neurol* 46:224-233.
- Peden AH, Ironside JW. (2004). Review: pathology of variant Creutzfeldt-Jakob disease. *Folia Neuropathol*; 42 (suppl A): 85-91.
- Petersen S, Bomme C, Baastrup C, Kemp A, Christoffersen GRJ (2002). Differential effects of mGluR<sub>1</sub> and mGluR<sub>5</sub> antagonism on spatial learning in rats. *Pharmacol Biochem Behav* 73:381-389.
- Phillis JW, O'Regan MH (2004). A potentially critical role of phospholipases in central nervous system ischemic, traumatic, and neurodegenerative disorders. *Brain Res Rev* 44:13-47.
- Pin JP, Acher F (2002). The metabotropic glutamate receptors: structure, activation, mechanisms and pharmacology. *Curr Drug Target CNS Neural Disord* 1:297-317.
- Pozas E., J. Ballabriga, A.M. Planas, I. Ferrer (1997). Kainic acid-induced excitotoxicity is associated with a complex c-Fos and c-Jun response which does not preclude either cell death or survival, *J. Neurobiol.* 33; 232-246.
- Prinster SC, Hague C, Hall RA (2005). Heterodimerization of G protein-coupled receptors: specificity and functional significance. *Pharmacol Rev* 57: 289-298.
- Prusiner SB. (1982). Novel proteinaceous infectious particles cause scrapie. *Science* 216:136-44.
- Prusiner SB (1997). The prion diseases of humans and animals. In: *The molecular and genetic basis of neurological diseases* (Rosenber RN, Prusiner SB, DiMauro S, Barchi RL, eds), pp 165-186. Boston: Butterworth-Heinemann.
- Prusiner SB (1998). Prions. *Proc Natl Acad Sci USA*; 95: 13363-83.

- Prusiner S.B., Groth, D., Serban, A. et al. (1993). Ablation of the prion protein (PrP) gene in mice prevents *scrapie* and facilitates production of anti-PrP antibodies. Proc. Natl. Acad. Sci. USA, 90, 10608-10612.
- Ralevic V, Burnstock G. (1998). Receptors for purines and pyrimidines. Pharmacol Rev; 50:413-92.
- Rebecchi MJ, Pentylala SN (2000). Structure, function, and control of phosphoinositidespecific phospholipase C. Physiol Rev 80: 1291-1335.
- Reppert SM, Weaver DR, Stehle JH, et al. (1991). Molecular cloning and characterization of a rat A1-adenosine receptor that is widely expressed in brain and spinal cord. Mol Endocrinol; 5:1037-48.
- Rhee SG (2001). Regulation of phosphoinositide-specific phospholipase C. Annu Rev Biochem 70:281-312.
- Richardson EP, Masters CL. (1995). The nosology of Creutzfeldt-Jakob disease and conditions related to the accumulation of PrP<sup>Sc</sup> in the nervous system. Brain Pathol. 5:33-4.
- Riemer C, Neidhold S, Burwinkel M, Schwartz A, Schultz J, Krätzschmar J, Mönning U, Baier M (2004). Gene expression profiling of *scrapie*-infected brain tissue. Biochem Biophys Res Commun 323:556-564.
- Riemer C, Queck I, Simon D, Kurth R, Baier M (2000). Identification of upregulated genes in *scrapie*-infected brain tissue. J Virol 74:10245-10248.
- Robinson M.J., M.H. Cobb (1997). Mitogen-activated protein kinase pathways, Curr. Opin. Cell Biol. 9; 180-186.
- Rodrigues SM, Bauer EP, Farb CR, Schafe GE, LeDoux JE (2002). The group I metabotropic glutamate receptor mGluR5 is required for fear memory formation and long-term potentiation in the lateral amygdala. J Neurosci 22:5219-5229.
- Rodríguez A, Freixes M, Dalfó E, et al. (2005). Metabotropic glutamate receptor/phospholipase C pathway: a vulnerable target to

- Creutzfeldt-Jakob disease in the cerebral cortex. *Neuroscience*; 131:825-32.
- Roy-Byrne PP (2005). The GABA-benzodiazepine receptor complex: structure, function, and role in anxiety. *J Clin Psychiatry* 66 Suppl 2:14-20.
  - Rudolphi KA, Shubert P. (1997). Modulation of neuronal and glial cell function by adenosine and neuroprotection in vascular dementia. *Behav Brain Res*; 83:123-8.
  - Saadoun S, Papadopoulos MC, Davies DC, Bell BA, Krishna S (2002). Increased aquaporin 1 water channel expression in human brain tumours. *Br J Cancer* 87:621-623.
  - Saadoun S, Papadopoulos MC, Davies DC, Krishna S, Bell BA (2002). Aquaporin-4 expression is increased in oedematous human brain tumours. *J Neurol Neurosurg Psychiatr* 72:262-265.
  - Sakudo A, Lee D, Saeki K, Nakamura Y, Inoue K, Matsumoto Y, et al. (2003). Impairment of superoxide dismutase activation by N-terminally truncated prion protein (PrP) in PrP-deficient neuronal cell line. *Biochem Biophys Res Commun*; 308:660-7.
  - Sakudo A, Lee D, Yoshimura E, Nagasaka S, Nitta K, Saeki K, Matsumoto Y, Lehmann S, Itohara S, Sakaguchi S, Onodera T (2004). Prion protein suppress perturbation of cellular copper homeostasis under oxidative conditions. *Biochem Biophys Res Commun* 313: 850-855
  - Sanz O., A. Estrada, I. Ferrer, A.M. Planas (1997). Differential cellular distribution and dynamics of HSP70, cyclooxygenase-2, and c-Fos in the rat brain after transient focal ischemia or kainic acid, *Neuroscience* 80; 221-232.
  - Sasaki N, Takeuchi M, Chowei H, Kikuchi S, Hayashi Y, Nakano N, et al. (2002). Advanced glycation end products (AGE) and their receptor (RAGE) in the brain of patients with Creutzfeldt-Jakob disease with prion plaques. *Neurosci Lett*; 326:117-20.

- Sato S, Umenishi F, Inamasu G, Sato M, Ishikawa M, Nishizawa M, Oizumi T (2000). Expression of water channel mRNA following cerebral ischemia. *Acta Neurochir Suppl* 76:239-241.
- Schnecko A, Witte K, Bohl J, et al. (1994). Adenylyl cyclase activity in Alzheimer's disease brain: stimulatory and inhibitory signal transduction pathways are differentially affected. *Brain Res*; 644:291-6.
- Schubert P, Ogata T, Ferroni S, et al. (1997). Protective mechanisms of adenosine in neurons and glial cells. *Ann NY Acad Sci*; 825:1-10.
- Schwartz M (2003). *How the cows turned mad*. Berkeley: University of California Pres.
- Sisó S, Puig B, varea R, Vidal E, Acín C, Prinz M, Montrasio F, Badiola J, Aguzzi A, Pumarola M, Ferrer I (2002). Abnormal synaptic protein expression and cell death in murine *scrapie*. *Acta Neuropathol* 103: 615-626.
- Smeyne R.J., M. Vendrell, M. Hayward, S.J. Baker, C.G. Miao, K. Schilling, L.M. Robertson, T. Carrant, J.I. Morgan (1993). Continuous c-fos expression precedes programmed cell death in vivo, *Nature* 363; 166-169.
- Solenov EI, Vetrivel L, Oshio K, Manley GT, Verkman AS (2002). Optical measurement of swelling and water transport in spinal cord slices from aquaporin null mice. *J Neurosci Meth* 113:85-90.
- Stern K. (1939). Severe dementia associated with bilateral symmetrical degeneration of the thalamus. *Brain* 62:157-71.
- Sun MC, Honey CR, Berk C, Wong NL, Tsui JK (2003). Regulation of aquaporin-4 in a traumatic brain injury model in rats. *J Neurosurg* 98:565-569.
- Taccola G, Marchetti C, Nistri A (2004). Modulation of rhythmic patterns and cumulative depolarization by group I metabotropic glutamate receptors in the neonatal rats cord in vitro. *Eur J Neurosci* 19:533-541.
- Taniguchi M, Yamashita T, Kumura E, Tamatani M, Kobayashi A, Yokawa T, Maruno M, Kato A, Ohnishi T, Kohmura E, Tohyama M,

- Yoshimine T (2000). Induction of aquaporin-4 water channel mRNA after focal cerebral ischemia in rat. *Mol Brain Res* 78:131-137.
- Taylor DR, Hooper M. (2006). The prion protein and lipid rafts. *Review. Mol Membr Biol*; 23:89-99.
  - Telling G.C., M. Scott, K.K. Hsiao, et al. (1994). Transmission of ECJ disease from humans to transgenic mice expressing chimeric human-mouse prion protein. *Proc. Natl. Acad. Sci. USA* Vol. 91, pp. 9936-9940.
  - Tsuboi Y, Baba Y, Don-ura K, Imamura A, Fujioka S, Yamada T (2005). DiVusion-weighted MRI in familial Creutzfeldt- Jakob disease with the codon 200 mutation in the prion protein gene. *J Neurol Sci* 232:45-49.
  - Ukisu R, Kushihashi T, Kitanosono T, Fujisawa H, Takenaka H, Ohgiya Y, Gokan T, Munechika H (2005). Serial diVusionweighted MRI of Creutzfeldt-Jakob disease. *Am J Roentgenol* 184:560-56655.
  - Vajda Z, Pedersen M, Fuchtbauer EM, Wertz K, Stodkilde-Jorgensen H, Sulyok E, Dóczy T, Neely JD, Agre P, Frokiaer J, Nielsen S (2002). Delayed onset of brain edema and mislocalization of aquaporin-4 in dystrophin-null transgenic mice. *Proc Natl Acad Sci USA* 99:13131-13136.
  - Vajda Z, Promeneur D, Dóczy T, Sulyok E, Frokiaer J, Ottersen OP, Nielsen S (2000). Increased aquaporin immunoreactivity in rat brain in response to systemic hyponatremia. *Biochem Biophys Res Commun* 270:495-503.
  - Van Everbroeck B, Dobbeleir I, De Waele M, de Leenheir E, Lübke U, Martin J-J, et al. (2004). Extracellular protein deposition correlatos with glial activation and oxidative stress in Creutzfeldt-Jakob and Alzheimer's disease. *Acta Neuropathol*; 108:194-200.
  - Venero JL, Machado A, Cano J (2004). Importance of aquaporins in the physiopathology of brain edema. *Curr Phar Des* 10:2153-2161.
  - Verkman AS (2002). Physiological importance of aquaporin water channels. *Ann Med* 34:192-200.

- Wakabayashi K, Narisawa-Saito M, Iwakura Y, Arai T, Ikeda K, Takahashi H, Nawa H (1999). Phenotypic down-regulation of glutamate receptor subunit GluR1 in Alzheimer's disease. *Neurobiol Aging* 20:287-295.
- Walton M.R., I. Dragunow (2000). Is CREB a key to neuronal survival?, *Trends Neurosci.* 23; 48-53.
- Wardas J. (2002). Neuroprotective role of adenosine in the CNS. *Pol J Pharmacol*; 54:313-6.
- Warth A, Mittelbronn M, Wolburg H (2005). Redistribution of the water channel protein aquaporin-4 and the K<sup>+</sup> channel protein Kir4.1 diVers in low- and high-grade human brain tumors. *Acta Neuropathol* 109:418-426.
- Wettschureck N, Offermanns S (2005). Mammalian G proteins and their cell type specific functions. *Physiol Rev* 85: 1159-1204.
- White AR, Collins SJ, Maher F, Jobling MF, Stewart LR, Thyer JM, et al. (1999). Prion protein-deficient neurons reveal lower glutathione reductase activity and increased susceptibility to hydrogen peroxide toxicity. *Am J Pathol*; 155:1723-30.
- Will RG, Ironside JW, Zeidler M, Cousens SN, Estibeiro K, Alperovitch A, et al. (1996). A new variant of Creutzfeldt-Jakob disease in the UK. *Lancet*; 347:921-925.
- Will RG, Zeidler M, Stewart GE, et al. (2000). Diagnosis of new variant Creutzfeldt-Jakob disease. *Ann Neurol*; 47: 575-82.
- Wittendorp MC, Von Frijtag Drabbe Künzel J, Ijzerman AP, et al. (2004). The mouse brain adenosine A1 receptor: functional expression and pharmacology. *Eur J Pharmacol*; 487:73-9.
- Wong BS, Liu T, Li R, Pan T, Petersen RB, Smith MA, et al. (2001). Increased levels of oxidative stress markers detected in the brains of mice devoid of prion protein. *J Neurochem*; 76:565-72.
- Wong BS, Pan T, Liu T, Li R, Petersen RB, Jones IM, et al. (2000). Prion disease: a loss of antioxidant functions? *Biochem Biophys Res Commun*; 275:249-52.

- Xiang W, Windl O, Westner IM, Neumann M, Zerr I, Lederer RM, Kretzschmar HA (2005). Cerebral gene expression profiles in sporadic Creutzfeldt-Jakob disease. *Ann Neurol* 58:242-257.
- Xiang W, Windl O, Wunsch, Dugas M, Kohlmann A, Dierkes N, Westner IM, Kretzschmar HA (2004). Identification of differentially expressed genes in *scrapie*-infected mouse brains by using global gene expression technology. *J Virol* 78:11051-11060.
- Xiao F, Arnold TC, Zhang S, Brown C, Alexander JS, Carden DL, Conrad SA (2004). Cerebral cortical aquaporin-4 expression in brain edema following cardiac arrest in rats. *Acad Emerg Med* 11:1001-1007.
- Yaar R, Jones MR, Chen J-F, et al. (2005). Animal models for the study of adenosine receptor function. *J Cell Physiol*; 202:9-20.
- Yamamoto, G.A. Gonzalez, W.H. Biggs, M.R. Montminy (1998). Phosphorylation induced binding and transcriptional efficacy of nuclear factor CREB, *Nature* 334; 494-498.
- Yamamoto M, Gotz ME, Ozawa H, et al. (2000). Hippocampal level of neural specific adenylyl cyclase type I is decreased in Alzheimer's disease. *Biochem Biophys Acta*; 1535: 60-8.
- Yamamoto M, Ozawa H, Saito T, Frolich L, Riederer P, Takahata N (1996). Reduced immunoreactivity of adenylyl cyclase in dementia of Alzheimer type. *Neuroreport* 7:2965-2970.
- Yamamoto-Sasaki M., H. Ozawa, T. Saito, M. Rosier, P. Riederer (1999). Impaired phosphorylation of cyclic AMP response element binding protein in the hippocampus of dementia of the Alzheimer type, *Brain Res.* 824; 300-303.
- Ye X., H.C. Meeker, P. Kozlowski, R.I. Carp (2002). Increased c-Fos protein in the brains of *scrapie*-infected SAMP8, SAMR1, AKR and C57BL mice, *Neuropathol. Appl. Neurobiol.* 28; 358-366.
- Zalewska-Kaszubska J. (2002). Neuroprotective mechanisms of adenosine action in CNS neurons. *Neurol Neurochir Pol*; 36:329-36.



