

## Prof. Adriano Aguzzi – MD, PhD hc, DVM hc, FRCP, FRCPath

Born December 1, 1960  
Italian citizen  
Permanent resident of Switzerland  
Married to Monika Landert, MD  
Two daughters, Chiara, born 21.10.2004,  
and Laura, born 18.6.2008



### *Currently held positions:*

- Professor of Neuropathology (Faculties of Medicine and of Science)
- Chairman, Department of Pathology
- Director, Institute of Neuropathology
- Director, Swiss National Reference Center for Prion diseases

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## Education and Training

INSTITUTION AND LOCATION	DEGREE (if applicable)	YEAR(s)	FIELD OF STUDY
University of Freiburg (D). Internship in Basel (CH) and Freiburg (D).	M.D.	1980-86	Medical School
Columbia University, New York, Dept. of Pathology)		1983	Immunology
Foreign medical graduates Examination in the Medical Sciences USA	ECFMG	1985	Human Medicine
University Hospital of Zürich, CH		1986-89	Resident in Neuropathology
Institute of Molecular Pathology, Vienna, A		1989-92	Postdoctoral training in molecular biology

## Positions

- 1993 Consultant (Oberarzt) and Lecturer (Privat-Dozent) in Pathology and Neuropathology, University of Zürich
- 1995 Director of the Swiss National Reference Center for Prion Diseases
- 1997 Ordinarius (Full Professor) of Neuropathology and Director of the Institute of Neuropathology, University of Zürich.
- 1998 Joint Professor of the Medical Faculty and Faculty of Natural Sciences, University of Zurich.
- 2004 Chairman, Department of Pathology, University Hospital of Zurich.

## Professional Memberships

- *Swiss Society of Neuropathology* (President: 1998-2002)
- *Royal College of Pathology* (FRCPath) since 1996, Fellow since 2003
- *German Society of Neuropathology*
- *International Society of Neurovirology* (Member of the Board of Directors 1999-2007)
- *International Society of Neuropathology* (Councilor 1997-99)
- *Spongiform Encephalopathy Advisory Committee* to the British Government (1997-2004)
- *Swiss Academy of the Medical Sciences* (SAMW): individual member since 2000
- *Academic Association "Forschung für Leben"* (President since 1999)
- *Italian Society of Virology*: Honorary Member
- *Austrian Academy of Sciences* since 2001: Corresponding member
- *German Academy of Sciences Leopoldina*: Co-opted member since 2001
- *American Society for Microbiology* (since 2000)
- *European Dana Alliance for the Brain, London*
- *Foederatio Medicorum Helvetiae*
- *Medical Society of Zurich*

## Honors

*Doctor medicinae honoris causa*, University of Liège, 2002

*Doctor medicinae veterinariae honoris causa*, University of Teramo, I, 2005

*Doctor scientiae honoris causa*, University of Bologna, 2002

*Fellow, Royal College of physicians (FRCP)*, London 2002

*German Academy of Science (Leopoldina) Prize*, Halle 2001

*Medal of the Royal Swedish Academy of Medicine*, Stockholm 2001

*ICAAC Award and Lecture*, Chicago 2001

*Aschoff Medal*, Freiburg 2000

*Cameron Lecturer*, Birmingham 2000

*Robert-Koch-Prize*, Berlin 2003

*Glaxo Award for Translational Neuroscience*, Lisbon 2004

*Emil von Behring Lecture*, Erfurt 2005

*Biotech Award*, Milano 2000

*Ernst-Jung Prize for Medicine*, Hamburg 1999

*Tibor Greenwalt Lecturer*, San Francisco 1999

*EMBO Gold Medal* of the European Molecular Biology Organization, Lisbon 1998

Membership to the *European Molecular Biology Organization*, 1998

*Cloëtta Award*, Zürich 1998

*Pfizer Award for Neurobiology*, Zürich 1997

*Ernst Th. Jucker Prize*, Zürich 1995

*Marcel-Benoist Prize*, Zurich 2004

*Sheikh Hamdan Bin Rashid Al Maktoum Award for Medical Sciences*, Dubai 2004

*Ernst Friedheim Lecture*, Rockefeller University, New York 2005

## Editorial activities

- Member of the Editorial Board of the following journals:

- *Acta Neuropathol.*
- *Brain Pathology*
- *Cell Death Different. (2002-2006)*
- *European Neurol. (until 2007)*
- *J. Exp. Pathol. ('96-'03)*
- *J. Neurovirology*
- *Neuropathol. Appl. Neurobiol.*
- *Pathologica*
- *Transgenics*
- *Virchows Arch. Pathol.*
- *Haematologica (until 2007)*
- *Glia (1998-2003)*
- *Int. J. Cancer (2000-2003)*
- *Trends in Microbiol.*
- *Molecular Aspects of Medicine*
- *Neuromol. Medicine*
- *The Lancet Neurol (2001-03)*
- *J. Mol. Neurosci.*
- *Journal of Neurology*
- *The EMBO Journal*
- *EMBO Reports*
- *Neurodegen. Diseases*
- *Nature Reviews Microbiology*
- *PLoS Medicine*
- *Nature Clin. Pract. Neurol.*
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- *Brain Pathology*: Deputy editor 1994-95, now Member of the Editorial Board.
- *Nature, Nature Medicine, Lancet*: referee on a regular basis
- *Science, Nature Genetics, J. Biochem., J. Mol. Biol., Neuroscience, Eur. Journal of Neurosci.*: ad hoc referee

## Extracurricular activities

- *President, Board of the Neuropath Foundation (since 2006)*
- *Chairman, Scientific Advisory Board, Institut Suisse de Recherche Expérimentale sur le Cancer, Epalinges, Switzerland (since 2002)*
- *Chairman, Scientific Advisory Board, Synapsis Foundation, Zurich, CH*
- *Director, Board of the European Brain Research Institute (since 2003)*
- *Program Director, MD-PhD program, University of Zürich (since 2004)*
- *Member of the following Scientific Advisory Boards:*
  - *Center for Molecular Biology ZMBH, Heidelberg, D (since 2001)*
  - *Wellcome Trust Neurosciences Committee, UK (2004-5)*
  - *Interdisciplinary Center for Neurosciences, Heidelberg, D*
  - *Julius-Klaus Foundation, Zürich*
  - *Charles-Rodolphe Brupbacher Foundation, Zürich (since 1998)*
  - *European Mouse Mutant Archive, Monterotondo, I*
  - *Giovanni Armenise-Harvard Foundation, Boston MA (until June 2008)*
- *Board of Governors (ETH-Rat), Swiss Federal School of Technology (ETH) (2003-2007)*
- *Board of Referees, Human Frontiers Science Program (1997-2001)*
- *Board of Directors, Roche Research Foundation (since 2003)*
- *Patronage committees:*
  - *Swiss National M.D.-Ph.D. program, Zürich*
  - *Postgraduate Program of the Universities of Zürich and Basel*
  - *Gen-Suisse Foundation, Berne*
  - *Sonnweid Alzheimer Foundation, Wetzikon CH*
- *Board of Trustees, International Forum for TSE and Food Safety, Berne CH*
- *Board of Trustees, Walter and Gertrud Siegenthaler Foundation*
- *TSE/BSE ad hoc Group, European commission, Brussels (1997-2002)*
- *Technical Steering Committee, Functional Genomics Center Zürich*
- *Prions Study Group, International Committee on Taxonomy of Viruses (1999 – 2002)*
- *Swiss Federal Committee for Biological Safety (2000-2001)*
- *Associate Dean for Research, University of Zurich Medical School (2000-2002)*
- *Visiting Professor at the Institute of Psychiatry, King's College, London UK (2000-2002)*
- *Scientific Advisory Board, Istituto di Ricerche Biomediche, Bellinzona (since 2005)*

## Patents

- *Aguzzi, A., Klein, M.A., Raeber, A.J., Weissmann, C., Zinkernagel, R.: Diagnostics and therapeutics for transmissible spongiform encephalopathy and method for the manufacture of non-infective blood products and tissue derived products. European Patent Application 97122186.6-1270 / WO9930738A2. Filed Dec. 8, 1997*
- *Aguzzi A., Fischer M.B. : Prion-binding activity in serum and plasma. Filed Sept. 28, 1999*
- *Aguzzi A., Genoud N., Räber A.J.: Soluble hybrid prion proteins and their use in the diagnosis, prevention, and treatment of transmissible spongiform encephalopathies. EP27008-03117. Filed March 14<sup>th</sup>, 2003*
- *Aguzzi, A.; Miele G.: Surrogate marker for early diagnostic of prion disease. Filed July 7<sup>th</sup>, 2004*

## **Research sketch:**

In 1992, Charles Weissmann asked me to collaborate with him and Hansruedi Büeler on the characterization of mice deficient in the normal prion protein. These mice turned out to be resistant to prions (1). I then found that expression of the normal prion protein by neurons is necessary for the development of histological damage (2). My lab showed that "neuroinvasion", *i.e.* the process by which prions march through the body of the host towards the brain, relies on expression of the normal prion protein in a non-hematopoietic extracerebral site (3). We then developed the hypothesis that neuroinvasion takes place in distinct steps: first the epithelial is trespassed (4), then the lymphoreticular system is colonized. Finally infectivity progresses from lymphoreticular organs to the central nervous system (5) via sympathetic nerves (6, 7). We are now testing the role of microglia using a conditional microglial paralysis model (8).

My lab set out to identify the lymphoreticular compartments responsible for the spread of prions. B-lymphocytes are required for peripheral prion pathogenesis (9), although the presence of the normal prion protein on B-lymphocytes is not necessary for this process (10). With Charles Weissmann, we found that one mechanism of action of B cells consists of presentation of lymphotoxins to follicular dendritic cells (FDCs) (11). This finding may pave the way to post-exposure prophylaxis (12) exploiting the anti-prion effect of soluble lymphotoxin- $\beta$  receptors. The distance between FDCs and splenic nerves was found to determine the velocity of neuroinvasion (13). M. Glatzel and I also found PrP<sup>Sc</sup> also in lymphoid organs of patients with Creutzfeldt-Jakob disease (14), suggesting similar pathogenetic mechanisms in humans and mouse models. As B-cells home to inflamed organs, we tested whether inflammation specifies the organ tropism of prions. This prediction proved correct (15).

But why do follicular dendritic cells accumulate prions? We tested whether prion uptake may be complement-mediated. Indeed, certain components of the complement system proved to play an important role in pathogenesis (16). Progress in the understanding of pathogenesis and a elucidation of possible therapeutical and prophylactic approaches, such as immunotherapy (17) must go hand-in-hand with diagnostic procedures. With this in mind, we initiated a screen for plasma proteins that bind specifically disease-associated prion protein (18, 19). In addition, we found that soluble dimeric prion protein binds selectively PrP<sup>Sc</sup> (20). This may be exploitable as the basis of a sensitive diagnostic assay, possibly for BSE and for human prion diseases.

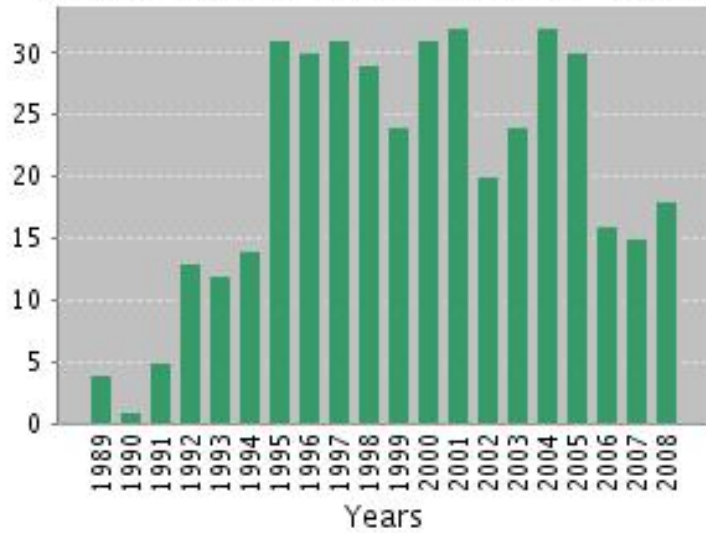
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2. Brandner, S., Isenmann, S., Raeber, A., Fischer, M., Sailer, A., Kobayashi, Y., Marino, S., Weissmann, C., and Aguzzi, A. 1996. Normal host prion protein necessary for scrapie-induced neurotoxicity. *Nature* 379:339-343.
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4. Heppner, F.L., Christ, A.D., Klein, M.A., Prinz, M., Fried, M., Kraehenbuhl, J.P., and Aguzzi, A. 2001. Transepithelial prion transport by M cells. *Nat Med* 7:976-977.
5. Aguzzi, A., and Weissmann, C. 1997. Prion research: the next frontiers. *Nature* 389:795-798.
6. Glatzel, M., Flechsig, E., Navarro, B., Klein, M.A., Paterna, J.C., Bueler, H., and Aguzzi, A. 2000. Adenoviral and adeno-associated viral transfer of genes to the peripheral nervous system. *Proc Natl Acad Sci U S A* 97:442-447.
7. Glatzel, M., Heppner, F.L., Albers, K.M., and Aguzzi, A. 2001. Sympathetic innervation of lymphoreticular organs is rate limiting for prion neuroinvasion. *Neuron* 31:25-34.
8. Heppner, F.L., Greter, M., Marino, D., Falsig, J., Raivich, G., Hovelmeyer, N., Waisman, A., Rulicke, T., Prinz, M., Priller, J., et al. 2005. Experimental autoimmune encephalomyelitis repressed by microglial paralysis. *Nat Med* 11:146-152.
9. Klein, M.A., Frigg, R., Flechsig, E., Raeber, A.J., Kalinke, U., Bluethmann, H., Bootz, F., Suter, M., Zinkernagel, R.M., and Aguzzi, A. 1997. A crucial role for B cells in neuroinvasive scrapie. *Nature* 390:687-690.

10. Klein, M.A., Frigg, R., Raeber, A.J., Flechsig, E., Hegyi, I., Zinkernagel, R.M., Weissmann, C., and Aguzzi, A. 1998. PrP expression in B lymphocytes is not required for prion neuroinvasion. *Nat Med* 4:1429-1433.
11. Montrasio, F., Frigg, R., Glatzel, M., Klein, M.A., Mackay, F., Aguzzi, A., and Weissmann, C. 2000. Impaired prion replication in spleens of mice lacking functional follicular dendritic cells. *Science* 288:1257-1259.
12. Aguzzi, A., and Collinge, J. 1997. Post-exposure prophylaxis after accidental prion inoculation. *Lancet* 350:1519-1520.
13. Prinz, M., Heikenwalder, M., Junt, T., Schwarz, P., Glatzel, M., Heppner, F.L., Fu, Y.X., Lipp, M., and Aguzzi, A. 2003. Positioning of follicular dendritic cells within the spleen controls prion neuroinvasion. *Nature* 425:957-962.
14. Glatzel, M., Abela, E., Maissen, M., and Aguzzi, A. 2003. Extraneural pathologic prion protein in sporadic Creutzfeldt-Jakob disease. *N Engl J Med* 349:1812-1820.
15. Heikenwalder, M., Zeller, N., Seeger, H., Prinz, M., Klohn, P.C., Schwarz, P., Ruddle, N.H., Weissmann, C., and Aguzzi, A. 2005. Chronic lymphocytic inflammation specifies the organ tropism of prions. *Science* 307:1107-1110.
16. Klein, M.A., Kaeser, P.S., Schwarz, P., Weyd, H., Xenarios, I., Zinkernagel, R.M., Carroll, M.C., Verbeek, J.S., Botto, M., Walport, M.J., et al. 2001. Complement facilitates early prion pathogenesis. *Nat Med* 7:488-492.
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18. Fischer, M.B., Roeckl, C., Parizek, P., Schwarz, H.P., and Aguzzi, A. 2000. Binding of disease-associated prion protein to plasminogen. *Nature* 408:479-483.
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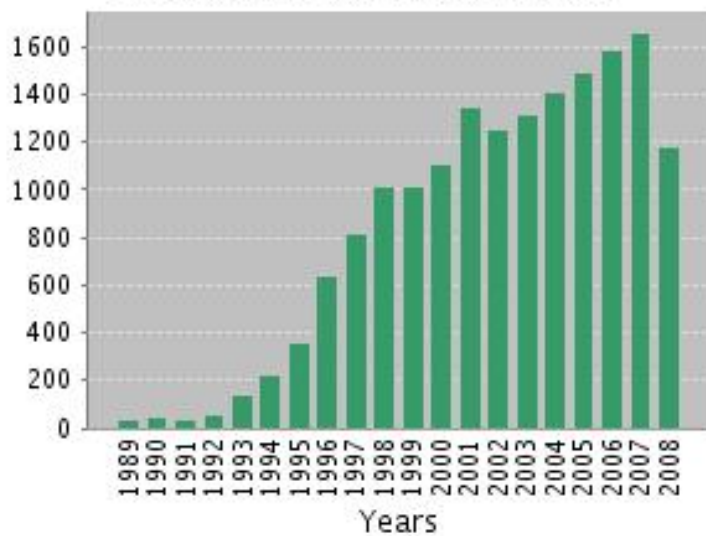
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Original papers

1. Giacomini, P., **Aguzzi, A.**, Pestka, S., Fisher, P.B. and Ferrone, S. (1984). Modulation by recombinant DNA leukocyte (alpha) and fibroblast (beta) interferons of the expression and the shedding of HLA- and tumor-associated antigens by human melanoma cells. *J. Immunol.* 133, 1649-1655.
2. Giacomini, P., Imberti, L., **Aguzzi, A.**, Fisher, P.B., Trinchieri, G. and Ferrone, S. (1985). Immunochemical analysis of the modulation of human melanoma-associated antigens by DNA recombinant immune interferon. *J. Immunol.* 135, 2887-2894.
3. Giacomini, P., **Aguzzi, A.**, Tecce, R., Fisher, P.B. and Ferrone, S. (1985). A third polypeptide associated with heavy and light chain subunits of class I HLA antigens in immune interferon treated human melanoma cells. *Eur. J. Immunol.* 15, 946-951.
4. Giacomini, P., **Aguzzi, A.** and Ferrone, S. (1985). Differential susceptibility to modulation by recombinant immune interferon of HLA-DR and -DQ antigens synthesized by melanoma COLO 38 cells. *Hybridoma* 5, 277-288.
5. Burger, P.C., Shibata, T., **Aguzzi, A.** and Kleihues, P. (1988). Selective induction by *N*-nitrosoethylurea of oligodendrogliomas in fetal forebrain transplants. *Cancer Res.* 48, 2871-2875.
6. Grant, J.W., Steart, P.V., **Aguzzi, A.**, Jones, D.B. and Gallagher, P.J. (1989). Gliosarcoma: an immunohistochemical study. *Acta Neuropathol. Berlin* 790, 305-309.
7. \* Williams, R.L., Risau, W., Zerwes, H.-G., Drexler, H., **Aguzzi, A.** and Wagner, E.F. (1989). Endothelioma cells expressing polyoma middle T oncogene induce hemangiomas by host cell recruitment. *Cell* 57, 1053-1063.
8. **Aguzzi, A.**, Wagner, E.F., Williams, R.L. and Courtneidge, S.A. (1990). Sympathetic hyperplasia and neuroblastomas in transgenic mice expressing polyoma middle T antigen. *The New Biologist* 2, 533-543.
9. **Aguzzi, A.**, Kleihues, P., Heckl, K. and Wiestler, O.D. (1991). Cell-type specific tumor induction by oncogenes in fetal forebrains transplants. *Oncogene*, 6, 113-118
10. Khazaie, K., Panayotou, G., **Aguzzi, A.**, Samarut, J., Gazzolo, L. and Jurdic, P. (1991). EGF promotes *in vivo* tumorigenic growth of primary chicken embryo fibroblasts expressing *v-myc* and enhances *in vitro* transformation by the *v-erbA* oncogene. *Oncogene* 6, 21-28
11. Boulter, C.A., **Aguzzi, A.**, Williams, R.L., Wagner, E.F., Evans, M.J. and Beddington, R. (1991). Aberrant development and twinning induced by *v-src* in chimaeric mouse embryos. *Development* 111, 357-366
12. Bothe, K., **Aguzzi, A.**, Lassmann, H., Rethwilm, A. and Horak, I. (1991). Progressive encephalopathy and myopathy in transgenic mice expressing human foamy virus genes. *Science* 253, 533-535
13. **Aguzzi, A.**, Bothe, K., Anhauser, I., Horak, I., Rethwilm, A., and Wagner, E.F. (1992) Expression of human foamy virus is differentially regulated during development in transgenic mice. *The New Biologist* 4, 225-237
14. Brüstle, O., **Aguzzi, A.**, Talarico, D., Kleihues, P., and Wiestler, O.D. (1992) Angiogenic activity of the *k-fgf/hst* oncogene in neural transplants. *Oncogene* 7, 1177-1183
15. Ellmeier, W., **Aguzzi, A.**, Kleiner, E., Kurzbauer, R., and Weith, A. (1992). Mutually exclusive expression of a helix-loop-helix gene and *N-myc* in human neuroblastomas and in normal development. *EMBO J.* 7, 2563-2571
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17. Juillerat-Jeanneret, L., **Aguzzi, A.**, Wiestler, O.D., Darekar, P., and Janzer, R.C. (1992). Dexamethasone regulates the activity of enzymatic markers of cerebral endothelial cell lines. *In Vitro* 28, 537-543.
18. Adams, B., Dörfler, P., **Aguzzi, A.**, Kozmik, Z., Urbanek, P., Maurer-Fogy, I., and Busslinger, M. (1992). The Pax-5 gene encoding the transcription factor BSAP is expressed in the developing CNS, adult testis and in B-lymphocytes. *Genes Dev.* 6, 1589-1607.
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20. Boulter, C.A., **Aguzzi, A.**, Evans, M.J., and Affara, N. (1992). A chimaeric mouse model for autosomal-dominant polycystic kidney disease. *Contrib. Nephrol.* 97, 60-70.
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22. **Aguzzi, A.**, Wagner, E.F., Netzer, K.O., Anhauser, I., Bothe, K., and Rethwilm, A. (1993). Human foamy virus proteins accumulate in neurons and induce multinucleated giant cells in the brain of transgenic mice. *Am. J. Pathol.* 142, 1061-1072.
23. \* Büeler, 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
24. Thomas, J.E., **Aguzzi, A.**, Soriano, P., Wagner, E.F., and Brugge, J.S. (1993). Induction of tumor formation and cell transformation by polyoma middle T antigen in the absence of *Src*. *Oncogene* 8, 2521-2529
25. \* Hilberg, F., **Aguzzi, A.**, Howells, N. and Wagner, E.F. (1993). Impaired hepatogenesis and embryonic death in *c-jun* deleted mice. *Nature* 365, 179-181.
26. Kiessling, M., Stumm, G., Xie, Y., Herdegen, T., **Aguzzi, A.**, Bravo, R., and Gass, P. (1993). Differential transcription and translation of immediate early genes in the gerbil hippocampus after transient global ischemia *J. Cer. Blood Flow Metab.* 13, 914-924.
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28. Kiefer, F., Anhauser, I., Soriano, P., **Aguzzi, A.**, Courtneidge, S.A., and Wagner, E.F. (1994) Endothelial cell transformation by polyomavirus middle T antigen in mice lacking Src-related kinases. *Current Biol.* 4, 100-109
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30. \* Sailer, A., Büeler, H., Fischer, M., **Aguzzi, A.**, and Weissmann, C. (1994) No propagation of prions in mice devoid of PrP. *Cell* 77: 967-968
31. Constam, D.B., Schmid, P., **Aguzzi, A.**, Schachner, M., and Fontana, A. (1994). Transient production of TGF- $\beta$ 2 by postnatal cerebellar neurons and its effects on neuroblast proliferation, aggregation and L1 adhesion molecule expression. *Eur. J. Neurosci.* 6, 766-778
32. McClure, M.O., Bienasz, P.D., Schulz, T.F., Chrystie, I.L., Simpson, G., **Aguzzi, A.**, Hoad, J.G., Cunningham, A., Kirkwood, J., and Weiss, R.A. (1994). Isolation of a new foamy virus from orangutans. *J. Virol.* 68, 7124-7130.

33. Magyar, J.P., Bartsch, U., Wang, Z.Q., **Aguzzi, A.**, Wagner, E.F., and Schachner, M. (1994) Degeneration of neural cells in the central nervous system of mice deficient in the gene for the adhesion molecule on glia. *J. Cell Biol.* 127, 135-146.
34. Günther, T., Struwe, M., **Aguzzi, A.**, and Schughart, K. (1994) Open brain, a new mouse mutant with severe neural tube defects, shows altered gene expression patterns in the developing spinal cord. *Development* 120, 3119-3123
35. **Aguzzi, A.**, and Theuring, F. (1994) An improved *in situ*  $\beta$ -galactosidase staining for histological analysis of transgenic mice. *Histochemistry* 102, 477-481.
36. Müller, U., Cristina, N., Li, Z-W., Wolfer, D.P., Lipp, H.P., Rüllicke, T., Brandner, S., **Aguzzi, A.**, and Weissmann, C. (1994) Behavioral and anatomical deficits in mice homozygous for a modified  $\beta$ -amyloid precursor protein ( $\beta$ -APP) gene. *Cell* 79, 755-766.
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38. Kiess, M., Scharm, B., **Aguzzi, A.**, Hajnal, A., Schwarte-Waldhoff, I., and Schäfer, R. (1995) Expression of *ril*, a novel LIM domain gene, is down-regulated in HRAS-transformed cells and restored in phenotypic revertants. *Oncogene* 10, 61-68
39. Stuart, E.T., Kioussi, C., **Aguzzi, A.**, and Gruss, P. (1995) PAX 5 proto-oncogene activity in human astrocytic tumours. *Clin. Cancer Res.* 1, 207-214
40. Hergersberg, M., Matsuo, K., Gassmann, M., Schaffner, W., Lüscher, B., Rüllicke, T., and **Aguzzi, A.** (1995) Tissue-specific expression of a FMR-1/ $\beta$ -galactosidase fusion gene in transgenic mice. *Hum. Mol. Genetics* 4, 359-366
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