

Curriculum Vitae

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Date of Birth: 19th May 1956 Nationality: British

Education:

1976-1979 BSc. 1st Class Hons. Genetics. Sheffield University
1982-1984 MSc. Biomolecular Organisation, London University
1984-1987 Ph.D. Molecular Genetics of Cystic Fibrosis, London University

Employment:

1979-1983	Clinical Cytogeneticist	Queen Elizabeth Hospital for Sick Children, London.
1983-1984	Research Assistant	St. Mary's Hospital Medical School, London.
1987-1993	Postdoctoral Fellow	Genome Analysis Laboratory, ICRF.
1994-1998	Senior Lecturer in Molecular Biology	Division of Medical and Molecular Genetics, United Medical and Dental Schools, London
1998-present	Professor of Neurogenetics	Dept. of Medical and Molecular Genetics, King's College London School of Medicine

Awards, Honours and Prizes

1993 National Medical Research Award of the National Health Council
1994 Liebermann Award, Hereditary Disease Foundation
1997 Principal Investigator HDSA Coalition for the Cure
1997 Liebermann Award, Hereditary Disease Foundation
1998 HDSA Milton Wexler Award for Research into HD
1998 Royal Society Glaxo Wellcome Award (with Stephen Davies)
1999 Max Planck Research Award for International Cooperation
1999 Member of the Academy of Medical Sciences
2000 Pius XI Medal Award for 1998 in recognition of outstanding research.
2001 Klaus Joachim Zülch-Preis
2002 Membership of EMBO (European Molecular Biology Organisation)

Membership of Boards, Committees.

1997-2001 Scientific Advisory Board of the Hereditary Disease Foundation
1998-2003 Editorial Board of Human Molecular Genetics
1999-2002 Associate Editor, Brain Pathology
2000-2003 Neuroscience Panel, Wellcome Trust

2001– present	Research Council, Huntington's Society Canada
2002-2007	Scientific Advisory Board of the Hereditary Disease Foundation
2003	Chair, Gordon Research Conference on CAG Triplet Repeat Diseases.
2003	Subcommittee to assess quinquennial review of MRC Prion Unit
2003 - present	Board of Directors of the Euro HD Network.
2003 - 2008	Guy's Committee for the Ethical Review Process in KCL.
2004 - present	HighQ-Psychogenics Advisory Committee
2006 – present	Editorial Advisory Board of New England Journal of Medicine
2006 – 2008	International Advisory Board for EU POLYALA project
2006 – 2009	Sectional Committee of the Academy of Medical Sciences

Membership of Professional Societies

1990 - present	The Genetical Society
1993 - present	World Federation of Neurology
1995 - present	The Human Genome Organisation
1996 - present	British Society for Human Genetics

Teaching

1989	EMBO course organiser - Long range mapping and cloning of mammalian chromosomes.
1994 - 2004	GKT School of Medicine 2nd year BMS medical genetics course. Lectures and tutorials.
1994 - present	KCL School of Medicine Intercalated BSc in Genetics. Neurogenetics course organiser.

Lectures given on the following courses:

1990 - 1996	MSc Medical Genetics, St. Mary's Hospital Medical School.
1991 - 1998	MSc Neurodegenerative Disease, University College London.
1995 - 1998	BSc Biochemistry, Intercollegiate.
1998 - 1999	Anatomy and Human Biology, GKT School of Medicine
1993 - 2001	MSc Medical Genetics with Immunology, Brunel University.
1998 - 2000	BSc Biochemistry, Imperial College.
1995 - present	BSc Molecular and Cellular Neuroscience, GKT School of Medicine
1995 - present	BSc Developmental Neurobiology, GKT School of Medicine
1999 - present	BSc Neuropharmacology and Neurotoxicity, GKT School of Medicine
2000	Molecular Mechanisms of Human Neurological Disease, Cold Spring Harbor, USA.
2000 - present	BSc, Neurogenetics module, St. Georges' Hospital Medical School.

Appointed Examiner

2001 - 2005	Chief External Examiner, MSc in Medical Genetics with Immunology, Brunel University
2002 - 2006	External Examiner, MSc in Clinical Neuroscience, Institute of Neurology, UCL.

PhD Students

Awarded 1995	Sarah Baxendale, ICRF
Awarded 2002	Kirupa Sathasivam, KCL (part time)
Awarded 2002	Caroline Benn, KCL (MRC studentship)
Awarded 2003	Donna Smith KCL (KCL studentship)
Awarded 2004	David Hay, KCL (MRC studentship)
Awarded 2006	Roman Gonitel (MRC studentship)
2004 – present	John Bett (Wellcome Prize Studentship)
2005 – present	Eva Sirinathsinghji (KCL, Harris Scholarship)

Grants Awarded

1. Hereditary Disease Foundation (Sole applicant)
The use of yeast artificial chromosomes to generate animals transgenic for the Huntington's disease gene. 1st August 1994 - 31st July 1996. £61,538 (\$100,000).
2. Medical Research Council (Sole applicant) Ref: E9330458
The use of yeast artificial chromosomes to generate animals transgenic for the Huntington's disease gene. 1st September 1994 - 31st August 1997. £176,657.
3. Medical Research Council (Co-applicant) Ref: G95067792MB
Identification of the gene for cystinosis by positional cloning. 1st October 1995 - 31st September 1998. £177,645.
4. National Kidney Research Fund (Co-applicant)
Identification of markers for the genetic analysis of Cystinosis. 1st March 1996 - 31st February 1998. £64,664.
5. Special Trustees of Guy's Hospital (Sole applicant)
Establishment and maintenance of colonies of mice transgenic for the human Huntington's disease mutation. 1st April 1996 - 31st March 1997. £28,709.
6. European Community (Principal applicant) Ref: BMH4 CT96 0244
Molecular and Clinical Neuropathology of Huntington's disease and Spinocerebellar Ataxia. 1st May 1996 - 31st April 1999. 410 kECU.
7. The Wellcome Trust (Principal applicant) Ref: 047852
Application for an ABI 377 Prism DNA sequencer. 1st June 1996 - 31st May 1999. £120,259.
8. Hereditary Disease Foundation (Sole applicant)
Maintenance and expansion of colonies of mice transgenic for the HD CAG expansion and entire mutant HD gene. 1st July 1996-31st March 1998. \$83,425.
9. Special Trustees of St. Thomas's Hospital (Sole applicant)
Expression analysis of the Fugu homologue of the HD gene in mouse cells, and its use for generating transgenic mice. 1st August 1996- 31st July 1997. £40,000.
10. The Wellcome Trust (Sole applicant) Ref: 000782
Characterisation of the mouse hap1 gene and an evaluation of its role in the progressive neurological phenotype observed in mice transgenic for the Huntington's disease mutation. 1st May 1997 - 31st April 2000. £187,155.
11. Hereditary Disease Foundation (Sole applicant)
Transgenic Approaches Toward an Understanding of the Molecular Basis of Huntington's Disease. 1st August 1997 - 31st July 1999. \$100,000.
12. The Huntington's Disease Society of America (Principal applicant)
Analysis of neuronal intranuclear inclusions in HD transgenic mice. 1st September 1997 - 31st August 1998. \$55,000.
13. The Wellcome Trust (Sole applicant) Ref: 051897
Toward an understanding of the molecular basis of Huntington's disease and the development of therapeutic interventions. 1st September 1997 - 31st August 2002. £863,357.

14. The Special Trustees of Guy's Hospital (Sole applicant)
Huntington's disease transgenic mouse colonies: management and assessment. 1st August 1998 - 31st July 1999. £18,179.
15. The Huntington's Disease Society of America (Principal applicant)
Molecular and ultrastructural characterisation of neuronal intranuclear inclusions (NII) in Huntington's disease. 1st October 1998 - 31st August 1999. \$50,000.
16. The Wellcome Trust (Co-applicant) Ref: 052780
Neuropathological characterisation of mouse models of Huntington's Disease. 1st October 1998 - 30th September 2001. £262,203.
17. Medical Research Council (Co-operative grant) Ref: G9800001
Genetic Approaches to Human Disease. 1st March 1999 - 28th February 2003. £249,012.
18. The Wellcome Trust (Co-applicant) Ref: 057036
Refurbishment and Equipment for ES Cell culture facility and IVC racks for mice. 1st June 1999 - 30th September 2002. £95,777.
19. The Huntington's Disease Society of America (Sole applicant)
Development of an organotypic slice culture assay as a screen to test the ability of various compounds and exogenously expressed proteins to prevent or delay the polyglutamine aggregation process. 1st October 1999 - 30th September 2000. \$75,000.
20. Human Frontiers Science Programme (Principal applicant) Ref: RG0132/1999-B
Prevention of polyglutamine aggregation - a therapeutic approach to Huntington's disease. 1st November 1999 - 31st October 2002 \$750,000.
21. Medical Research Council JREI (Co-applicant) Ref: G9900846
Microarray technology in studies of neurodegeneration, immunology, development and oncogenesis. Commencing January-2000 £52,290.
24. Max Planck Society Award (Sole awardee) Ref: V-2 MPF-GRO/1064161
The molecular pathogenesis of Huntington's disease and the development of therapeutic strategies. January 2000 - December 2004. DM 250,000.
22. Wellcome Trust - Equipment Grant (Principal applicant) Ref: 060360
To establish a facility dedicated to the production of transgenic and gene targeted mouse lines and the breeding and analysis of mouse models with a wide variety of phenotypes and pathogen containment requirements. June 2000 - May 2003. £674,205.
23. KCL Postgraduate Research Studentship
Development of an organotypic slice culture assay from Huntington's disease transgenic mice for screening polyglutamine aggregation inhibitors. Sept. 2000 - August 2003 £39,565.
24. The Huntington's Disease Society of America (Sole applicant)
Slice culture assay to assess approaches aimed at modifying polyglutamine aggregation in HD and the characterisation of transgenic models designed to dissect the molecular basis of the phenotype. 1st October 2000 - 30th September 2002. \$191,194.
25. The Wellcome Trust - Joint Infrastructure Fund (Co-applicant) Ref: 990430-06
Refurbishment of the Division of Medical and Molecular Genetics. £6,400,000

26. Medical Research Council JREI (Co-applicant) Refs: JR00KCANEQ/JR00KCANRC
Protein mass spectrometry in biomedical research
January 2001 - December 2003. £294,539
27. The Wellcome Trust – Program grant (Sole Applicant) Ref. 066270
Approaches to Understand the Molecular Basis of Huntington's disease and Develop Therapeutic Strategies. 1st September 2002 – 31st August 2007. £1,578,241
28. The Hereditary Disease Foundation (Principal Applicant)
Experimental therapeutics in HD mouse models.
1st Feb 2003 – 30th June 2005. \$482,349
29. The Huntington's Disease Society of America (Sole applicant)
Pharmacological and Genetic Approaches to Modifying the Phenotype in Mouse Models of HD.
1st January 2003 - 31st December 2003. \$108,197.
30. Wellcome Trust Prize Studentship Ref: 073061
Impairment of the ubiquitin proteasome pathway in Huntington's disease pathogenesis.
January 1st 2004 – 31st December 2006. £106,339
31. The Hereditary Disease Foundation (Sole applicant)
Analysis of transcriptional changes in HD mouse models – request for funds to purchase a real time PCR machine.
September 2003. £24,645.75
32. The Huntington's Disease Society of America (Principal Applicant)
Folding aggregation and clearance of mutant huntingtin.
January 1st 2004 – 31st December 2006. \$330,000.
33. The High Q Foundatoin (Elaine Holmes PI Sarah Tabrizi and Gill Bates co-applicants)
Metabonomic strategy for the identification of biomarkers of Huntington's disease in mouse models and HD populations.
October 1st 2004 – September 30th 2006. £138,527
34. CHDI Inc. / HighQ Foundation. (Bates and Lowden)
Preclinical assessment of novel therapeutic compounds in mouse models of HD.
October 1st 2005 – September 30th 2007. £346,457
35. King's College London Studentship (Harris Scholarship).
Generation of a human stem cell model of HD.
October 1st 2005 – September 30th 2008. £90,000
36. Hereditary Disease Foundation (Bates and Lowden)
Analytical and synthetic chemistry to support the preclinical testing of compounds in mouse models of HD.
January 1st 2006 – December 31st 2007. \$189,378.58
37. Huntington's Disease Association (UK)
Generation and characterization of a human embryonic stem cell model of HD
October 1st 2006 – September 30th 2009. £43,800

Patent Applications

Patent number: US2005227915

Publication date: 13th October 2005.

Title: Methods and reagents for treating neurodegenerative disorders and motor deficit disorders.

Inventors: Steffan JS (US), Thompson LM (US), Marsh JL (US), Bodai L (US), Pallos J (US), Hockly E (GB), Bates GP (GB).

Patent number: DE69834193D

Publication date: 25th May 2006

Title: Novel method of detecting amyloid-like fibrils or protein aggregates

Inventors: Wanker E (DE), Lehrach H (DE), Scherzinger E (DE), Bates G (GB).

Patent number: US7078191

Publication date: 18th July 2006

Title: Composition and methods for the detection of disease associated amyloid-like fibril or protein aggregate formation.

Inventors: Wanker E (DE), Lehrach H (DE), Scherzinger E (DE), Bates G (GB).

Publications - Gillian Bates

- Bates GP (2006). One misfolded protein allows others to sneak by. *Science* 311, 1385-1386.
- Bates GP, Gonitel R (2006). Mouse models of triplet repeat diseases. *Molecular Biotechnology*, 32, 147-158.
- Bett J, Bates GP, Hockly E. (2006). Molecular Pathogenesis and Therapeutic Strategies in Huntington's disease. Eds. Well R and Ashizawa T. In *Genetic Instabilities and Neurological Diseases*, 2nd Ed. Elsevier Press, CA, pp 223 - 249.
- Bett JS, Goellner GM, Woodman B, Pratt G, Rechsteiner M, Bates GP.(2006) Proteasome impairment does not contribute to pathogenesis in R6/2 Huntington's disease mice: exclusion of proteasome activator REG γ as a therapeutic target. *Hum. Mol. Genet.* 15, 33-44.
- Borkqvist M, Petersen A, Bacos K, Isaacs J, Norlen P, Gil J, Popovic N, Sundler F, Bates GP, Tabrizi SJ, Brundin P, Mulder H (2006). Progressive alterations in the hypothalamic-pituitary-adrenal axis in the R6/2 transgenic mouse models of Huntington's disease. *Hum. Mol. Genet.* 15, 1713-1721.
- Butler R, Bates GP, (2006). HDAC inhibitors as therapeutics for polyglutamine disease. *Nature Reviews Neuroscience*. in press.
- Hockly E, Tse J, Barker AL, Moolman DL, Beunard J-L, Revington AP, Holt K, Sunshine S, McPhail H, Sathasivam K, Woodman B, Wanker EE, Lowden PAS, Bates GP (2006). Evaluation of benzothiazole aggregation inhibitors riluzole and PGL-135 as therapeutics for Huntington's disease. *Neurobiol. Dis.* 21, 228-236.
- Guidetti P, Bates GP, Graham RK, Hayden MR, Leavitt BR, MacDonald ME, Slow EJ, Wheeler VC, Woodman B, Schwarcz R (2006). Elevated brain 3-hydroxykynurenine and quinolinate levels in Huntington's disease mice. *Neurobiol. Dis.* In press.
- Tarditi A, Camurri A, Varani K, Borea P-A, Woodman B, Bates G, Cattaneo E, Abbracchio MP (2006). Early and transient alteration of adenosine A_{2A} receptor signaling in a mouse model of Huntington's disease. *Neurobiol. Dis.* In press.
- Tsang TM, Woodman B, McGloughlin G, Griffin JL, Tabrizi SJ, Bates GP, Holmes E (2006). Metabolic characterisation of the R6/2 transgenic mouse model of Huntington's disease by high-resolution MAS ¹H NMR spectroscopy. *J. Proteome. Res.* 5, 483-492.
- Woodman B, Butler R, Landles C, Lupton MK, Tse J, Hockly E, Moffitt H, Sathasivam K, Bates GP (2006). The *Hdh*^{Q150/Q150} knock-in mouse model of HD and the R6/2 exon 1 model develop comparable and widespread molecular phenotypes. *Brain Research Bulletin*, in press.
- Bates GP (2005). The molecular genetics of Huntington's disease – a history. *Nature Reviews Genetics*, 6, 766-773.
- Benn CL, Landles C, Li H, Strand AD, Woodman B, Sathasivam K, Li S-H, Ghazi-Noori S, Hockly E, Faruque SMNN, Cha J-J, Sharpe PT, Olson JM, Li X-J, Bates GP (2005). Contribution of nuclear and extranuclear polyQ to neurological phenotypes in mouse models of Huntington's disease. *Hum. Mol. Genet.* 14, 3065-3078.
- Cornett J, Cao F, Wang C-E, Ross C, Bates GP, Li S-H, Li X-J (2005). Polyglutamine expansion of huntingtin impairs its nuclear export. *Nat. Genet.* 37, 198-204.

- Henley SM, Bates GP, Tabrizi SJ (2005). Biomarkers for neurodegenerative disease. *Curr. Opin. Neurol.* 18, 698-705.
- Miller TW, Zhou C, Gines S, MacDonald ME, Mazarakis ND, Bates GP, Huston JS, Messer A (2005). A human single-chain Fv intrabody preferentially targets amino-terminal huntingtin fragments in striatal models of Huntington's disease. *Neurobiol. Dis.* 19, 47-56.
- Papalexi E, Persson A, Bjorkqvist M, Petersen A, Woodman B, Bates GP, Sundler F, Mulder H, Brundin P, Popovic, N (2005). Reduction of GnRH and fertility in the R6/2 mouse model of Huntington's disease. *Eur. J. Neuroscience* 22, 1541-1546.
- Smith R, Petersen A, Bates GP, Brundin P, Li J-Y (2005). Depletion of rabphilin 3A in a transgenic mouse model (R6/1) of Huntington's disease, a possible culprit in synaptic dysfunction. *Neurobiol. Dis.* 19, 47-56.
- Valenza M, Rigamonti D, Goffredo D, Zuccato C, Fenu S, Jamot L, Strand A, Tarditi A, Woodman B, Racchi M, Mariotti C, DiDonato S, Corsini A, Bates G, Pruss R, Olson J, Sipione S, Tartari M and Cattaneo E (2005). Dysfunction of the cholesterol biosynthetic pathway in Huntington's disease. *J. Neurosci.* 25, 9932-9939.
- Zhang X, Smith DL, Meriin AB, Engemann S, Russel DE, Roark M, Washington SL, Marsh JL, Thompson LM, Wanker EE, Young AB, Housman DE, Bates GP, Sherman MY, Kazantsev AG (2005). A potent small molecule inhibits polyglutamine aggregation in vitro and suppresses neurodegeneration in vivo. *Proc. Natl. Acad. Sci.* 102, 892-897.
- Bates GP, Hay DG. (2004) Mouse models of triplet repeat diseases. *Trinucleotide Repeat Protocols. Methods in Molecular Biology* 277, 3-15. Ed. Yoshinori Kohwi. Humana Press, Totowa New Jersey.
- Hay DG, Sathasivam S, Tobaben S, Stahl, B, Marber M, Mestrlil R, Mahal, A, Smith DL, Woodman, B, Bates GP. (2004) Progressive decrease in chaperone protein levels in a mouse model of Huntington's disease and induction of the stress response as a therapeutic approach. *Hum. Mol. Genet.* 13: 1389-1405.
- Landles C, Bates GP (2004) Huntington and the molecular pathogenesis of Huntington's disease. *EMBO reports* 5: 958-963.
- Smith DL, Bates GP. (2004) Monitoring aggregate formation in organotypic slice cultures from transgenic mice. *Methods in Molecular Biology* 277, 161-171. Ed. Yoshinori Kohwi. Humana Press, Totowa New Jersey.
- Bates GP (2003) Huntington aggregation and toxicity in Huntington's disease. *Lancet*, 361: 1642-1648.
- Bates GP and Hockly E. (2003) Experimental therapeutics in Huntington's disease - are models useful for therapeutic trials. *Curr. Op. Neurol.* 16, 465-470.
- Gourfinkel-An I, Parain K, Hartmann A, Mangiarini L, Brice A Bates GP, Hirsch EC (2003). Changes in GAD67 mRNA expression evidenced by in situ hybridization in the brain of R6/2 transgenic mice. *J. Neurochem.* 86, 1369-1378.
- Hockly, E, Richon VM, Woodman, B, Smith, DL, Zhou, X, Rosa, E, Sathasivam, K, Ghazi-Noori, S, Mahal, A, Lowden, PAS, Steffan, JS, Marsh, JL, Thompson, LM, Lewis, CM, Marks, PA, Bates, GP (2003). Suberoylanilide hydroxamic acid, a histone deacetylase inhibitor, ameliorates motor deficits in a mouse model of Huntington's disease. *Proc. Natl. Acad. Sci. USA*, 100, 2041-2046.
- Hockly, E, Woodman, B, Mahal, A, Lewis, CM, Bates GP (2003). Standardization and Statistical Approaches to Therapeutic Trials in the R6/2 mouse. *Brain Research Bulletin*, 61, 469-479.

- Hockly E, Beunard JL, Lowden P & Bates GP (2003) Minocycline is not beneficial in a phenotype mouse model of Huntington's disease. (Reply to a letter) *Ann Neurol*, 54 (6) 842-843.
- Luthi-Carter R, Apostol BL, Dunah AW, DeJohn MM, Farrell LA, Bates GP, Young AB, Standaert DG, Thompson LM, Cha J-HJ. (2003). Complex alterations of NMDA receptors in transgenic Huntington's disease mouse brain: analysis of mRNA and protein expression, plasma membrane association, interacting proteins and phosphorylation. *Neurobiol. Dis.* 14, 624-636.
- Orth M, Cooper JM, Bates GP, Schapira AH. (2003). Inclusion formation in Huntington's disease R6/2 mouse muscle cultures. *J. Neurochem.* 87, 1-6.
- Smith, DL, Woodman B, Mahal A, Sathasivam K, Ghazi-Noori S, Lowden PAS, Bates GP, Hockly E (2003). Minocycline and doxycycline do not improve phenotype in the R6/2 model of HD. *Annals Neurol.* 54, 186-196.
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- Bates GP and Murphy K (2002). Mouse models of Huntington's disease. In *Huntington's Disease 3rd ed.* Eds. Bates GP, Harper P and Jones L. Oxford University Press, UK, pp 387-426.
- Hay DG and Bates G (2002) Huntingtin. In *Wiley Encyclopaedia of Molecular Medicine*, 1681 - 1684. John Wiley and Sons, Inc.
- Hockly E, Cordery PM, Woodman B, Mahal A, van Dellen A, Blakemore C, Lewis S, Hannan AJ, Bates GP (2002) Environmental enrichment slows disease progression in R6/2 Huntington's disease mice. *Annals Neurol.* 51, 235-242.
- Lievens J-C, Woodman B, Mahal A, Bates, GP (2002). Abnormal phosphorylation of synapsin I predicts a neuronal transmission impairment in the R6/2 Huntington's disease transgenic mice. *Molecular and Cellular Neuroscience.* 20, 638-648.
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- Zabel C, Chamrad DC, Priller J, Woodman B, Meyer HE, Bates GP, Klose J (2002). Alterations in the mouse and human proteome caused by Huntington's disease. *Molecular and Cellular Proteomics*, 1, 366-375.
- Bates GP (2001). Huntington's disease – exploiting expression. *Nature* 413, 691-693.
- Hansson O, Castilho RF, Korhonen L, Linholm D, Bates GP, Brundin P (2001) Partial resistance to malonate-induced striatal cell death in transgenic mouse models of Huntington's disease is dependent on age and CAG repeat length. *J. Neurochem.* 78, 694-703.
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- Kusakabe M, Mangiarini L, Laywell ED, Bates GP, Yoshiki A, Hiraiwa N, Inoue J, Steindler DA. (2001) Loss of cortical and thalamic neuronal tenascin-C expression in a transgenic mouse expressing exon1 of the human Huntington disease gene. *Journal of Comparative Neurology* 430, (4) 485-500.

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- Smith DL, Portier R, Woodman B, Hockly E, Mahal A, Klunk WE, Li X-J, Wanker EE, Murray KD, Bates GP (2001) Inhibition of polyglutamine aggregation in R6/2 HD brain slices - complex dose response profiles. *Neurobiol. Dis.* 8, 1017-1026.
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- Bates GP, Eberwine J. (2000) Hunting in the calm before the storm. *Nature Genetics* 25, 365-366.
- Bates GP (2000) Huntington's disease - In reverse gear. *Nature* 404, 944-945.
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- Murphy KPSJ, Carter RJ, Lione LA, Mangiarini L, Mahal A, Bates GP, Dunnett SB, Morton AJ. (2000) Abnormal Synaptic Plasticity and Impaired Spatial Cognition in Mice Transgenic for Exon 1 of the Human Huntington's Disease Mutation. *J. Neurosci.* 20 (13), 5115-5123.
- Steffan JS, Kazanste A, Spasic-Boskovic O, Greenwald M, Zhu Y-Z, Gohler H, Wanker EE, Bates GP, Housman DE, Thompson LM (2000) The Huntington's disease protein interacts with p53 and CREB-binding protein and repress transcription. *Proc. Natl. Acad. Sci. USA*, 97(12) 6763-6768.
- Tabrizi SJ, Workman J, Hart P, Mangiarini L, Mahal A, Bates G, Cooper JM, Schapira AHV (2000) Mitochondrial dysfunction and free radical damage in the R6/2 transgenic mouse model of Huntington's disease. *Annals Neurol.* 47, 80-86
- Turmaine M, Raza A, Mahal A, Mangiarini L, Bates GP, Davies SW (2000) Nonapoptotic neurodegeneration in a transgenic mouse model of Huntington's disease. *PNAS.* 97 (14) 8093-8097.
- Bates GP, Mangiarini L, Davies SW (1999) Transgenic Mouse Models of Huntington's Disease; in *Central Nervous System Diseases*. Eds: DF Emerich, RL Dean and PR Sanberg, Humana Press Inc., Totowa, 335-367.
- Carter RJ, Lione LA, Humby T, Mangiarini L, Mahal, A, Bates GP, Morten AJ, Dunnett SB (1999) Characterisation of progressive motor deficits in mice transgenic for the human Huntington's disease mutation. *J. Neurosci.* 19 (8), 3248-3257.
- Cha J-H, Frey AS, Alsdorf SA, Kerner JA, Kosinski CM, Mangiarini L, Penney JB, Davies SW Bates GP Young AB (1999) Altered neurotransmitter receptor expression in transgenic mouse models of Huntington's disease. *Phil. Trans. Royal Soc.* 354, 981-989.
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- Davies SW, Turmaine M, Cozens BA, Mangiarini L, Bates GP (1999) From neuronal inclusions to neurodegeneration: neuropathological investigations of a transgenic mouse model of Huntington's disease. *Phil. Trans. Royal Soc.* 354, 971-979.

- Kosinski CM, Cha J-H, Young A, Mangiarini L, Bates GP, Schwarz M (1999) Intranuclear inclusions in subtypes of striatal neurons in Huntington's disease transgenic mice. *Neuroreport* 10, 3891-3896.
- Li H, Li S-H, Cheng AL, Mangiarini L, Bates GP, Li X-J (1999) Ultrastructural localisation and progressive formation of neuropil aggregates in Huntington disease transgenic mice. *Hum. Mol. Genet.* 8, 1227-1236.
- Lione LA, Rebecca JC, Hunt MJ, Bates GP, Morton AJ, Dunnet SB (1999) Selective discrimination learning impairments in mice expressing the Huntington's Disease Mutation. *J. Neurosci.* 19(23): 10428-10437.
- Mangiarini L, Sathasivam K, Bates GP (1999) Molecular pathology of Huntington's Disease: Animal models and nuclear mechanisms. *The Neuroscientist.* 5(6) 383-391.
- Reynolds GP, Dalton CF, Tillery CL, Mangiarini L, Davies SW, Bates GP (1999) Brain neurotransmitter deficits in mice transgenic for the Huntington's disease mutation. *J. Neurochem.* 72, 1773-1776.
- Sathasivam K, Hobbs C, Turmaine M, Mangiarini L, Mahal A, Bertaux F, Wanker EE, Doherty P, Davies SW, Bates GP (1999) Formation of polyglutamine inclusions in non-CNS tissue. *Hum. Mol. Genet.* 8, 813-822.
- Sathasivam K, Hobbs C, Mangiarini L, Mahal A, Turmaine M, Doherty P, Davies SW, Bates GP. (1999) Transgenic models of Huntington's disease. *Phil. Trans. Royal Soc.* 354, 963-969.
- Scherzinger E, Sittler A, Heiser V, Schweiger K, Hasenbank R, Bates GP, Lehrach H, Wanker EE (1999) Self-assembly of polyglutamine-containing huntingtin fragments into amyloid-like fibrils: implications for Huntington's disease pathology. *Proc. Natl., Acad. Sci. (USA)* 96, 4604-4609.
- Bates GP, Mangiarini L, Davies SW (1998) Transgenic mice in the study of polyglutamine repeat expansion diseases. *Brain Pathology*, 8, 699-714.
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