



## **NF Clinic Network (NFCN) Application Form\***

**Clinic Name:**

Washington University NF Clinic

**Affiliated Hospital:**

St. Louis Children's Hospital

**Affiliated University or Institution:**

Washington University

**Clinic Address:**

Washington University  
One Children's Place  
St. Louis, MO 63110

**Clinic Director:**

David H. Gutmann, MD, Ph.D.

**Clinic Coordinator Name:**

Anne C. Albers, RN, PNP

*\*Note: Some non-public information has been removed from this application form.*



## The Neurofibromatosis Clinic Network (NFCN)

### FORM PART A: Affiliate Clinic Application

#### 1. ABOUT YOUR NF CLINIC

a. Is your NF Clinic:

- Freestanding
- Hospital based
- In an academic center
- Other (please describe)

b. Describe overall your NF Clinic, when it meets and how it functions.

The Washington University NF Clinic meets twice monthly on the second and fourth Thursday beginning at 8am and concluding around 5p. Patients (adults and children) are seen by [Dr. David Gutmann] and the NF Clinic Coordinator, Anne Albers, PNP. We typically see ~16 patients per clinic. Additional weekly uncomplicated return patient clinics are being planned for later this fall. Concurrent with our clinic are our dedicated consultant clinics (ophthalmology, neuro-oncology, and dermatology) and select patients requiring these services are seen the same day. Radiologic consultations are available during clinic from our consultants in Neuroradiology.

## 2. CLINIC DIRECTOR and STAFF EXPERTISE

### a. CLINIC DIRECTOR: Please describe:

#### i. Your experience to date with NF care

*David H. Gutmann, MD, PhD has cared for patients with NF for the past 17 years, beginning as a fellow with Francis S. Collins, MD, PhD at the University of Michigan. Dr. Gutmann was recruited to Washington University to establish a Neurofibromatosis Clinical Program here. This program began seeing patients in 1994, and has grown to one of the largest experienced multi-disciplinary tertiary referral centers nationally. Another aspect of our commitment to NF clinical care is our strong relationship with the local NF chapter and its affiliates. Over the past decade, we have participated in numerous educational symposia, parent support group meetings, and community-based functions to raise awareness for NF in our region. Our integral role in the St. Louis community has been recognized over the years by the National Neurofibromatosis Foundation, including the National Neurofibromatosis Foundation (NFFF) Crystal Award in 1998 and the NFFF Center of Excellence Award in 2002.*

#### ii. Your past and current association with NF clinical trials

*Dr. Gutmann and his associates have participated in all available clinical trials for patients with NF, including the DOD-funded Natural History Study of Plexiform Neurofibromas in NF1 as well as several clinic trials involving Perfenidone (Allison King, MD), Farnesyltransferase (Allison King, MD), and AZD (Gerald Linette, MD, PhD).*

#### iii. Your past and current association with other clinical trials e.g. oncology trials

*Dr. Gutmann is a funded member of the newly established Department of Defense-sponsored NF Clinical Trials Consortium. He serves as the chair of the Biology Committee.*

### b. CLINIC DIRECTOR: Please provide information on:

#### i. Present and past funding you have received for NF research. Include funding source, date received, amount and project description.

*Clinical Investigator K08 Award, NINDS, 1992-1997*

*FIRST Award R29, NINDS, 1994-1999*

*Muscular Dystrophy Association, 1994-1996*

*National Tuberous Sclerosis Association, 1995-1999*

*Subcontract, NIH R01 (J. Lynn Rutkowski, P.I.), 1996-2000*

*NIH, R01 "Functional Analysis of NF2 Mutations," 1997-2000; Renewed 2000-2004*

*Subcontract, US Army Department of Defense, "Natural History of Plexiform neurofibromas in NF1" (Bruce Korf, PI), 1998-2002*

*Tuberous Sclerosis Alliance, Center without Walls, 1999-2004*

*NIH, R01 "Neurofibromin as a negative regulator for astrocytes", 1999-2003*

*NIH, P01, "ES Cell Transplantation After Spinal Cord Injury" (Dennis Choi, P.I.), 2000-2004*

American Cancer Society, "Preclinical Models of Human Astrocytomas", 2000-2003  
 NIH, R01 "Preclinical Mouse Models for Human Astrocytomas", 2001-2005  
 NIH, R01, "Protein 4.1 Tumor Suppressors in Meningioma Pathogenesis", 2001-2004  
 Co-PI, US Army Department of Defense, "Expression Profiling of Cell Lines Expressing Regulated NF2 Transcripts" (Stefan Pulst, PI), 2001-2003  
 Subcontract, US Army Department of Defense, "A Phase II Trial of the Farnesyltransferase inhibitor R115777 in Pediatric Patients with NF1" (Brigitte Widemann, PI), 2001-2004  
 Co-PI, US Army Department of Defense, "Development of mouse models of NF2-related meningiomas", (M. Giovannini, PI), 2002-2005  
 PI, US Army Department of Defense, "Mouse models of TSC-related epilepsy", 2003-2005  
 NIH, R01 "Gene Expression Analysis in Tuberous Sclerosis", 2003-2006  
 PI, US Army Department of Defense, "Modeling NF1-associated astrocytomas in vitro and in vivo", 2003-2007  
 PI, James S. McDonnell Foundation, "Mouse Models of Meningioma", 2003-2006  
 PI, James S. McDonnell Foundation, "Development of Preclinical Mouse Models of Meningioma", 2004-2006  
 PI, US Army Department of Defense, "Functional analysis of Protein 4.1 tumor suppressors", 2004-2008  
 Co-investigator, National Cancer Institute, Mouse Models of Human Cancers Consortium, "Astrocytic Cancers: How, When and Where?" (Terry Van Dyke, PI), 2004-2009  
 PI, US Army Department of Defense, "NF Consortium Site at Washington University", 2006-2012  
 PI, US Army Department of Defense, "Identification and Preclinical Evaluation of New Therapies for NF1 Brain Tumors", 2006-2010  
 PI, NIH, R21 "Identification and Preclinical Evaluation of New Brain Tumor Therapies", 2006-2008  
 PI, Brain Tumor Society, "Identification of Key Genetic and Growth Control Pathway Changes in JPA that Represent Potential Molecular Targets for Therapeutic Intervention", 2007  
 PI, Brain Tumor Society, "Identification of Key Genetic and Growth Control Pathway Changes in Pediatric Fibrillary Astrocytoma that Represent Potential Molecular Targets for Therapeutic Intervention", 2007-2008  
 PI, US Army Department of Defense, "Regulation and Role of mTOR in Neurofibromin Growth Control", 2007-2010  
 PI, NIH, R21 "Molecular Determinants of Neural Stem Cell Function", 2007-2009

- ii. Your NF-related clinical and scientific publications.  
 Include Journal, Citation and Title.

Peer-reviewed Manuscripts:

1. Groux N, Gutmann DH: Carcinoma of the lung. *Physician Assistant-Health Care Practitioner* 4:20-29, 1980.
2. Doneen BA, Gutmann DH: Lipid composition and in vitro biosynthetic rates of neutral lipids and phosphatidylcholine and anterior and posterior chambers of the goldfish swimbladder. *Comparative Biochemistry and Physiology A* 60:291-295, 1981.
3. Schook LB, Bingham EL, Gutmann DH, Niederhuber JE: Characterization and expression of H-2I region gene products on bone marrow derived macrophages. *European Journal of Immunology* 12:991-997, 1982.
4. Gutmann DH: The use of restriction endonucleases in the prenatal diagnosis of hemoglobinopathies. *American Journal of Medical Technology* 48:361-366, 1982.

5. Gutmann DH, Allen PM, Niederhuber JE: Separation of the immune response genes for LDH-B and MOPC-173. I. Description of an immune response defect in B10.BASR1. *J. Immunol.* 131:2919-2923, 1983.
6. Gutmann DH: Restriction Endonucleases. *American Journal of Medical Technology.* 49:60, 1983.
7. Gutmann DH, Niederhuber JE: Separation of the immune response genes for LDH-B and MOPC-173: II. Description of an immune response defect in BIO.ASR7. *J. Immunol.* 132:1955-1959, 1984.
8. Allen PM, Gutmann DH, Sher MR, Niederhuber JE: Characterization of the three new Intra-I-Region recombinant mouse strains, B10.ASR7 (H-2<sup>as3</sup>), B10.BAR4 (H-2<sup>h4</sup>), and B10.BASR1 (H-2<sup>as4</sup>). *Immunogenetics* 19:175-178, 1984.
9. Schook LB, Gutmann DH, Marlin LE, Niederhuber JE: In vitro derived bone marrow macrophages: Expression of Ia antigens during macrophage differentiation. *Transplantation* 37:585-590, 1984.
10. Gutmann DH, Niederhuber JE: Separation of the immune response genes for LDH-B and MOPC-173. III. The failure of B10. BASR1 to respond to LDH-B is due to an antigen presenting cell defect. *Transplantation*; 40:556-562, 1985.
11. Gutmann DH, Niederhuber JE: Major histocompatibility complex regulation of the immune response. *J. Surg Res.* 39:172-181, 1985.
12. Gutmann DH, Beard BA, Collinge ML: Nonfatal disseminated mucocutaneous herpes simplex virus type 2 infection in an otherwise healthy woman presenting with a urinary tract infection. *Obstetrics and Gynecology* 72:506-508, 1988.
13. Gutmann DH, Fischbeck KH, Sergott RC: Hereditary vasculopathy with cerebral white matter lesions. *Am J Med Genet* 34:217-220, 1989.
14. Gutmann DH, Scherer S: Ataxic hemiparesis syndrome: Localization to the corona radiata by magnetic resonance imaging. *Stroke* 20:1571-1573, 1989.
15. Raps EC, Gutmann DH, Brorson J, O'Connor MJ, Hurtig HH: Symptomatic hydrocephalus and reversible spinal cord deformity in *Listeria monocytogenes* meningitis. *J. Neurosurg.* 71:620-622, 1989.
16. Gutmann DH, Fischbeck KH: Molecular biology of Duchenne and Becker Muscular Dystrophy: Clinical Applications. *Ann Neurol* 26:189-194, 1989.
17. Gutmann DH, Fischbeck KH, Kamholz J: Complicated hereditary spastic paraparesis with cerebral white matter lesions. *Am J Med Genet* 36:251-7, 1990.
18. Gutmann DH, Grossman RI, Mollman JE: Personality changes associated with thalamic infiltration. *J. Neuro-oncology* 263-267, 1990.
19. Gutmann DH, Cantor CR, Piacente GJ, McCluskey LF: Isolated cerebral vasculopathy and infarction in a woman with carcinomatous meningitis. *J Neuro-oncology* 9:183-5, 1990.

20. Gutmann DH, Marino P: *An alternative apnea test for the evaluation of brain death. Annals of Neurology 30:852-853, 1991.*
21. Gutmann DH: *Chromosome 11q23.3-qter deletion and Alexander's disease. American Journal of Medical Genetics 39:226, 1991.*
22. Gutmann DH, Zackai EH, McDonald-McGinn DM, Fischbeck KH, Kamholz J: *Oculodentodigital dysplasia syndrome associated with abnormal cerebral white matter. Am J Medical Genetics 41:18-20, 1991.*
23. Gutmann DH, Brooks ML, Emmanuel BS, McDonald-McGinn DM, Zackai EH: *Congenital nystagmus in a (46XX, 45X) mosaic female from a family with X-linked congenital nystagmus. Am J Medical Genetics 39:167-169, 1991.*
24. Yousem DM, Gutmann DH, Milestone BN, Lenkinski RE: *Integrated magnetic resonance imaging and spectroscopy in a family with complicated hereditary spastic paraparesis. American J. Neuroradiology 12:785-790, 1991.*
25. Gutmann DH, Wood DL, Collins FS: *Identification of the neurofibromatosis gene type 1 product. Proceedings of The National Academy of Sciences, USA 88:9658-9662, 1991.*
26. Marchuk DA, Saulino AM, Tavakkol R, Swaroop M, Wallace MR, Andersen LB, Mitchell AL, Gutmann DH, Boguski M, Collins FS: *cDNA cloning of the type 1 neurofibromatosis gene: Complete sequence of the NF1 gene products. Genomics 11:931-940, 1991.*
27. Basu TN, Gutmann DH, Fletcher JA, Glover TW, Collins FS, Downward J: *Aberrent regulation of ras proteins in tumor cells from type 1 neurofibromatosis patients. Nature 356:713-715, 1992.*
28. Gutmann DH, Zackai EH, Emanuel BS: *Congenital nystagmus in a (46XX, 45X) mosaic female from a family with X-linked congenital nystagmus: a reply. Am. J. Med. Genetics 43:897, 1992.*
29. Gutmann DH, Collins FS: *Recent progress towards understanding the molecular biology of von Recklinghausen neurofibromatosis. Annals of Neurology 31:555-561, 1992.*
30. Gutmann DH, Boguski M, Marchuk D, Wigler M, Collins FS, Ballester R: *Analysis of the neurofibromatosis type 1 (NF1) GAP-related domain by site directed mutagenesis. Oncogene 8:761-769, 1993.*
31. Andersen LB, Ballester R, Marchuk DA, Chang E, Gutmann DH, Saulino AM, Camonis J, Wigler M, Collins FS: *A conserved alternative splice in the von Recklinghausen neurofibromatosis (NF1) gene produces two neurofibromin isoforms, both with GAP activity. Mol. and Cell. Biology 13:487-495, 1993.*
32. Gregory PE, Gutmann DH, Mitchell AL, Park S, Jacks T, Wood DL, Boguski M, Jove R, Collins FS: *The neurofibromatosis type 1 gene product co-localizes with microtubules. Somatic Cell and Molecular Genetics 13:487-495, 1993.*
33. Andersen LB, Fountain JW, Gutmann DH, Tarle SA, Glover TW, Dracopoli NC, Housman DE, Collins FS: *Mutations in the neurofibromatosis 1 gene in sporadic malignant melanoma. Nature Genetics 3:118-121, 1993.*

34. Gutmann DH, Andersen LB, Cole JL, Swaroop M, Collins FS: An alternatively-spliced mRNA in the carboxy terminus of the neurofibromatosis type 1 (NF1) gene is expressed in muscle. *Human Molecular Genetics* 2:989-992, 1993.
35. Gutmann DH, Tennekoon GI, Cole JL, Collins FS, Rutkowski JL: Modulation of the neurofibromatosis type 1 (NF1) gene product, neurofibromin, during Schwann cell differentiation. *J. Neuroscience Res.* 36:216-223, 1993.
36. Boyer M, Gutmann DH, Collins F, Bar-Sagi D: Co-capping of neurofibromin, but not GAP, with surface immunoglobulins in B lymphocytes. *Oncogene* 9:349-357, 1994.
37. Nakamura T, Nemoto T, Arai M, Yamazaki Y, Kasuga T, Gutmann DH, Collins FS, Ishikawa T: Specific expression of the neurofibromatosis type 1 (NF1) gene in the hamster Schwann cell. *Am. J. Path.* 144:549-555, 1994.
38. Gutmann DH, Cole JL, Stone WJ, Ponder BAJ, Collins FS: Loss of neurofibromin in adrenal gland tumors from patients with neurofibromatosis type 1. *Genes, Chromosomes and Cancer* 10:55-58, 1994.
39. Yousem DM, Gutmann DH, Lenkinski RE: Limitations of Magnetic Resonance Spectroscopy in patients with white matter disease. *Annals of Neurology* 36:932-933, 1994.
40. Gutmann DH, Cole JL, Collins FS: Modulation of neurofibromatosis type 1 (NF1) gene expression during in vitro myoblast differentiation. *J. Neuroscience Res.* 37:398-405, 1994.
41. Gutmann DH, Geist RT, Rose K, Wright DE: Expression of two new protein isoforms of the neurofibromatosis type 1 gene product, neurofibromin, in muscle tissues. *Dev. Dynamics* 202:302-311, 1995.
42. Gutmann DH, Silos-Santiago I, Geist RT, Daras M, Rutkowski JL: Lack of NF1 gene expression in a sporadic schwannoma from a patient without neurofibromatosis. *J. Neuro-Oncology* 25:103-111, 1995.
43. Gutmann DH, Geist RT, Rose K, Wallin G, Moley JF: Loss of neurofibromatosis type 1 (NF1) gene expression in pheochromocytomas from patients without NF1. *Genes, Chromosomes and Cancer* 13:104-109, 1995.
44. Gutmann DH, Geist RT, Wright DE, Snider WD: Expression of the neurofibromatosis type 1 (NF1) isoforms in rat embryonic and adult tissues. *Cell Growth and Differentiation* 6:315-322, 1995.
45. Norton KK, Carey JC, Gutmann DH: Oculodentodigital dysplasia with cerebral white matter lesions in a two generation family. *Am. J. Med. Genetics* 57:458-461, 1995.
46. Gutmann DH, Wright DE, Geist RT, Snider WD: Expression of the neurofibromatosis 2 (NF2) gene isoforms during rat embryonic development *Human Molecular Genetics* 4:471-478, 1995.
47. Norton KK, Xu J, Gutmann DH: Expression of the neurofibromatosis 1 gene product, neurofibromin, in blood vessel endothelial cells and smooth muscle. *Neurobiology of Disease* 2:13-21, 1995.

48. Hewett SJ, Choi DW, Gutmann DH: Increased expression of the neurofibromatosis 1 (NF1) tumor suppressor protein, neurofibromin, in reactive astrocytes in vitro. *NeuroReport* 6:1505-1508, 1995.
49. Geist RT, Gutmann DH: The tuberous sclerosis 2 gene is expressed at high levels in the adult cerebellum and developing spinal cord. *Cell Growth and Differentiation* 6:1477-1483, 1995.
50. Guha A, Lau N, Huvar I, Gutmann DH, Provias J, Pawson T, Boss G: Ras-GTP levels are elevated in human NF1 peripheral nerve tumors. *Oncogene* 12:507-513, 1996.
51. Giordano MJ, Mahadeo DK, He YY, Geist RT, Hsu C, Gutmann DH: Increased expression of the neurofibromatosis 1 (NF1) gene product, neurofibromin, in astrocytes in response to cerebral ischemia. *J. Neuroscience Res.* 43:246-253, 1996.
52. Feldkamp MM, Lau N, Provias JP, Gutmann DH, Guha A: Acute presentation of a neurogenic sarcoma in a patient with neurofibromatosis type 1: a pathological and molecular explanation. *J. Neurosurg.* 84:867-873, 1996.
53. Norton KK, Mahadeo DK, Geist RT, Gutmann DH: Expression of the neurofibromatosis 1 (NF1) tumor suppressor gene product, neurofibromin, during growth arrest in fibroblasts. *NeuroReport* 7:601-604, 1996.
54. Gutmann DH, Giordano MJ, Mahadeo DK, Lau N, Silbergeld D, Guha A: Increased neurofibromatosis 1 gene expression in astrocytic tumors: positive regulation by p21-ras. *Oncogene* 12:2121-2107, 1996.
55. Platten M, Giordano MJ, Dirvem CMF, Gutmann DH, Louis DN: Upregulation of specific NF1 gene transcripts in sporadic pilocytic astrocytomas. *Am. J. Pathol.* 149:621-627, 1996.
56. Geist RT, Gutmann DH: Expression of a developmentally-regulated neuron-specific isoform of the neurofibromatosis 1 (NF1) gene. *Neuroscience Letters* 211:85-88, 1996.
57. Geist RT, Reddy AJ, Zhang J, Gutmann DH: Expression of the tuberous sclerosis 2 (TSC2) gene product, tuberlin, in adult and developing nervous system tissues. *Neurobiology of Disease* 3:111-120, 1996.
58. Gurney JG, Shannon KM, Gutmann DH: Juvenile xanthogranuloma, neurofibromatosis 1, and juvenile chronic myeloid leukemia. *Archives of Dermatology* 132:1390, 1996.
59. McDonald JW, Bautista RE, Gutmann DH: Pseudo-cervical cord syndrome: a deceptive flumazenil-reversible manifestation of hepatic encephalopathy. *Archives of Neurology* 53:956, 1996.
60. Scherer SS, Gutmann DH: Expression of the neurofibromatosis 2 tumor suppressor gene product, merlin, in Schwann cells. *J. Neurosci. Res.* 46:595-605, 1996.
61. Gutmann DH: Tumor suppressor genes as negative growth regulators in development and differentiation. *International Journal of Developmental Biology* 39:895-907, 1996.
62. Listernick R, Louis DN, Packer RJ, Gutmann DH: Optic pathway gliomas in children with neurofibromatosis 1: consensus statement from the NF1 optic pathway glioma task force. *Annals of Neurology* 41:143-149, 1997.

63. Gutmann DH, Giordano MJ, Fishback AS, Guha A: Loss of merlin expression in sporadic meningiomas, ependymomas and schwannomas. *Neurology* 49:267-270, 1997.
64. Weinecke R, Guha A, Maize JC, Heideman RL, DeClue JE, Gutmann DH: Reduced TSC2 RNA and protein in sporadic astrocytomas and ependymomas. *Annals of Neurology* 42:230-235, 1997.
65. Xu HM and Gutmann DH: Mutations of the GAP-related domain impair the ability of neurofibromin to associate with microtubules. *Brain Research* 759:149-152, 1997.
66. Saporito-Irwin SM, Geist RT and Gutmann DH: Ammonium acetate protocol for the preparation of plasmid DNA suitable for mammalian cell transfections. *Biotechniques* 23:424-427, 1997.
67. Gutmann DH, Aylsworth A, Carey JC, Korf B, Marks J, Pyeritz RE, Rubenstein A, and Viskochil D: The diagnostic evaluation and multidisciplinary management of neurofibromatosis 1 and neurofibromatosis 2. *J. American Med. Assoc.* 278:51-57, 1997.
68. Gutmann DH, Saporito-Irwin S, DeClue JE, Weinecke R, and Guha A: Alterations in the rap1 signaling pathway are common in human gliomas. *Oncogene* 15:1611-1616, 1997.
69. Sherman L, Xu HM, Geist RT, Saporito-Irwin S, Howells N, Ponta H, Herrlich P and Gutmann DH: Interdomain binding mediates tumor growth suppression by the NF2 gene product. *Oncogene* 15:2505-2509, 1997.
70. Gutmann DH: Psychiatry and care of patients with neurofibromatosis. *J. American Med. Assoc.* 278:1494, 1997.
71. Gutmann DH: Molecular insights into neurofibromatosis 2. *Neurobiology of Disease* 3; 247-221, 1997.
72. Vogelbaum MA, Tong JX, Higashikubo R, Gutmann DH, Rich KM: Transfection of C6 glioma cells with the bax gene result in increased sensitivity to treatments with cytosine arabinoside. *J. Neurosurgery* 88:99-105, 1998.
73. Xu HM and Gutmann DH: Merlin differentially associates with the microtubule and actin cytoskeleton. *J. Neuroscience Research* 51:403-415, 1998.
74. Gutmann DH, Geist RT, Xu HM, Kim JS, and Saporito-Irwin S. Defects in neurofibromatosis 2 protein function can arise at multiple levels. *Human Molecular Genetics* 7:335-345, 1998.
75. Feldkamp MM, Gutmann DH, Guha A: Neurofibromatosis type 1: piecing the puzzle together. *Can. J. Neurosci.* 25:181-191, 1998.
76. Gutmann DH, Sherman L, Seftor L, Haipek C, Lu K-H, and Hendrix M: Increased expression of the NF2 suppressor gene product, merlin, impairs cell motility, adhesion and spreading. *Human Molecular Genetics* 8:267-276, 1999.
77. Strauss BL, Gutmann DH, Dehner LP, Hirbe A, Zhu X, Marley EF and Liapis H. Molecular analysis of malignant Triton tumors. *Human Pathology* 30:984-988, 1999.

78. Gutmann DH, Loehr A, Zhang Y, Kim J, Henkemeyer M and Cashen A: Haploinsufficiency for the neurofibromatosis 1 (NF1) tumor suppressor results in increased astrocyte proliferation. *Oncogene* 18:4450-4459, 1999.
79. Vogelbaum MA, Tong JX, Peroga R, Gutmann DH and Rich KM: Overexpression of bax in human glioma cell lines. *J. Neurosurg.* 91:483-489, 1999.
80. Hung G, Faudoa R, Li XK, Xeu Z, Brackmann DE, Hitselberg W, Saleh E, Lee F, Gutmann DH, Slattery WJ, Rhim JS, Lim D: Establishment of primary vestibular schwannoma cultures from neurofibromatosis type-2 patients. *International Journal of Oncology* 14:409-415, 1999.
81. Dugan LL, Kim JS, Zhang Y, Bart RD, Sun Y, Holtzman DM, Gutmann DH: Differential effects of cAMP in neurons and astrocytes - role of B-raf. *J. Biol. Chem.* 274:25842-25848, 1999.
82. Gutmann DH, Zhang Y, Hirbe A: Developmental regulation of a neuron-specific neurofibromatosis 1 (NF1) isoform. *Annals of Neurology* 46:777-782, 1999.
83. Liapis H, Dehner LP, Gutmann DH: Neurofibroma and cellular neurofibroma with atypia: a report of 14 tumors. *Am J. Surg. Pathol.* Sept. 23: 1156-1158, 1999.
84. Gutmann DH, Haipok CA, Lu KH: The neurofibromatosis 2 tumor suppressor protein, merlin, forms two functionally important intramolecular associations. *J. Neurosci. Res.* 58:706-716, 1999.
85. Listernick R, Charrow J, Gutmann DH: Intracranial gliomas in NF1. *Am. J. Med. Genet.* 89: 38-44, 1999.
86. Gutmann DH, Zhang Y, Hasbani MJ, Goldberg MP, Plank TL, Henske, EP: Expression of the tuberous sclerosis complex (TSC) gene products, hamartin and tuberin, in central nervous system tissues. *Acta Neuropathologica* 99:223-230, 2000.
87. JL Rutkowski, Wu K, Gutmann DH, Boyer P, Legius E: Multiple mechanisms of benign tumor formation in neurofibromatosis 1. *Human Molecular Genetics* 9:1056-1066, 2000.
88. Waggoner DJ, Towbin J, Gottesman G, Gutmann DH: A clinic-based study of plexiform neurofibromas in neurofibromatosis 1. *Am. J. Medical Genetics* 92:132-135, 2000.
89. King AA, DeBaun MR, Riccardi VM, Gutmann DH: Malignant peripheral nerve sheath tumors in neurofibromatosis 1. *Am. J. Medical Genetics* 93:388-392, 2000.
90. Gutmann DH, Donahoe J, Brown T, James CD, Perry A: Loss of neurofibromatosis 1 (NF1) gene expression in NF1-associated pilocytic astrocytomas. *Neuropath. Applied Neurobiol.* 26:361-367, 2000.
91. Gutmann DH, Donahoe J, Perry A, Lemke N, Gorse K, Kittiniyom K, Rempel SA, Gutierrez JA, Newsham IF: Loss of DAL-1, a Protein 4.1-related tumor suppressor, is an important early event in the pathogenesis of meningiomas. *Human Molecular Genetics* 9:1495-1500, 2000.
92. Lau N, Feldkamp MM, Roncari L, Loehr AH, Shannon P, Gutmann DH, Guha A: Loss of neurofibromin is associated with activation of ras/MAPK and PI3-K/Akt signaling in a neurofibromatosis 1 astrocytoma. *J. Neuropath Exp. Neurology* 59:759-767, 2000.

93. Scoles DR, Huynh DP, Chen MS, Burke SP, Gutmann DH, Pulst SM: The neurofibromatosis 2 (NF2) tumor suppressor protein interacts with hepatocyte growth factor-regulated tyrosine kinase substrate, HRS. *Human Molecular Genetics*. 9:1567-1574, 2000.
94. Perry A, Cai DX, Scheithauer BW, Swanson PE, Lohse CM, Newsham IR, Weaver A, Gutmann DH: Merlin, DAL-1 and progesterone receptor expression in clinicopathologic subsets of meningioma: A correlative immunohistochemical study of 175 cases. *J. Neuropath. Exp. Neurol.* 59:872-879, 2000.
95. Lim DJ, Rubenstein AE, Evans GE, Gusella JF, Jacks T, MacCollin M, Seizinger BG, Baser ME, Beebe D, Brackman DE, Chiocca EA, Fehon RG, Giovannini M, Glazer R, Gutmann DH, Korf B, Lieberman F, Martuza R, McClatchey AI, Parry DM, Pulst SM, Ramesh V, Ramsey J, Ratner N, Rutkowski JL, Rutledge M, Weinstein DE: Recent advances in neurofibromatosis type 2 (NF2): A workshop report. *J. Neurogenetics* 14:63-106, 2000.
96. Gutmann DH, Hirbe AC, Huang Z-y, Haipek CA: The Protein 4.1 tumor suppressor, DAL-1, impairs cell motility, but regulates proliferation in a cell type-specific fashion. *Neurobiol of Disease* 8:266-278, 2001.
97. Listernick R, Charrow J, Gutmann DH: Magnetic resonance findings and ophthalmologic abnormalities are correlated in patients with neurofibromatosis type 1 (NF1). (letter) *Am. J. Med. Genet.* 102:105, 2001.
98. Bajenaru ML, Donahoe J, Corral T, Reilly KM, Brophy S, Pellicer A, Gutmann DH: Neurofibromatosis 1 (NF1) Heterozygosity results in a cell-autonomous growth advantage for astrocytes. *GLIA* 33:314-323,2001.
99. Li J, Perry A, James CD, Gutmann DH: Cancer-related gene expression profiles in NF1-associated pilocytic astrocytomas. *Neurology* 56:885-890, 2001 (accompanying editorial; pages 827-829).
100. Morrison H, Sherman LS, Legg J, Banine F, Isacke C, Haipek CA, Gutmann DH, Ponta H, Herrlich P: The NF2 tumor suppressor gene product, merlin, mediates contact inhibition of growth through interactions with CD44. *Genes & Development* 15:968-980, 2001.
101. Gutmann DH, Haipek CA, Burke SP, Sun CX, Scoles DR, Pulst SM: The NF2 interactor, hepatocyte growth factor-regulated tyrosine kinase substrate (HRS), associates with merlin in the "open" conformation and suppresses cell growth and motility. *Human Molecular Genetics* 10:825-834, 2001.
102. Ding H, Roncari L, Shannon P, MacMaster S, Wu X, Lau N, Karaskova J, Gutmann DH, Squire JA, Nagy A, Guha A: Astrocyte-specific expression of activated p21-ras results in malignant astrocytoma formation in a transgenic mouse model of human gliomas1. *Cancer Research* 61:3826-3836, 2001.
103. Perry A, Roth KA, Banerjee R, Gutmann DH: NF1 deletions in S-100-positive and negative cells of sporadic and neurofibromatosis 1(NF1)-associated plexiform neurofibromas and MPNSTs. *Am. J. Pathol.* 159:57-61, 2001.
104. Scherer SS, Xu T, Crino P, Arroyo EJ, Gutmann DH: Ezrin, radixin and moesin are components of Schwann cell microvilli. *J. Neuroscience Res.* 65:150-164, 2001.

105. Gutmann DH, Hirbe AC, Haippek CA: *Functional analysis of neurofibromatosis 2 (NF2) missense mutations. Human Molecular Genetics 10:1519-1529, 2001.*
106. Li C, Cheng Y, Gutmann DH, Mangoura D: *Differential localization of the neurofibromatosis 1 (NF1) gene product, neurofibromin, with the F-actin or microtubule cytoskeleton during differentiation of telencephalic neurons. Developmental Brain Research 130:231-248, 2001.*
107. Perry A, Giannini C, Raghavan R, Scheithauer BW, Banerjee R, Margraf L, Bowers DC, Lytle RA, Newsham IF, Gutmann DH. *Aggressive Phenotypic and Genotypic Features in Pediatric and NF2-Associated Meningiomas: A Clinicopathologic Study of 53 Cases. J Neuropathol Exp Neurol. 60:994-1003, 2001.*
108. Kyin R, Hua Y, Baybis M, Scheithauer B, Kolson, D, Uhlmann EJ, Gutmann, DH, Crino, PB. *Differential Expression of Neurotrophins in Tuberous Sclerosis Complex-Associated Cortical Tubers. Am. J. Pathol. 159:1541-1554, 2001.*
109. Arbiser JL, Yeung R, Weiss SW, Arbiser ZK, Amin MB, Cohen C, Frank D, Mahajan S, Herron GS, Yang J, Onda H, Zhang HB, Bai X, Uhlmann E, Loehr A, Northrup H, Au P, Davis I, Fisher DE, Gutmann DH. *The generation and characterization of a cell line derived from a sporadic renal angiomyolipoma: use of telomerase to obtain stable populations of cells from benign neoplasms. Am J Pathol 159:483-491, 2001.*
110. Uhlmann EJ, Gutmann DH: *Tumor suppressor gene regulation of cell growth: Recent insights into neurofibromatosis 1 and 2 gene function. Cell Biochem and Biophysics 34:61-78, 2001.*
111. Reed N, Gutmann DH: *Tumorigenesis in neurofibromatosis: New insights & potential therapies. Trends in Molecular Medicine. 7:157-162, 2001.*
112. Gutmann DH, Wu YL, Hedrick NM, Zhu Y, Guha A, Parada LF: *Heterozygosity for the neurofibromatosis 1 (NF1) tumor suppressor results in abnormalities in cell attachment, spreading and motility in astrocytes. Human Molecular Genetics 10:3009-3016, 2001.*
113. Gutmann DH: *Neurofibromatosis: When less is more. Human Molecular Genetics. 10:747-755, 2001.*
114. MacCollin M, Gutmann DH, Korf B, Finkelstein R: *Establishing priorities in neurofibromatosis research: a workshop summary. Genet. Med. 3:212-217, 2001.*
115. Sherman LS, Gutmann DH: *Merlin: Hanging tumor suppression on the Rac. Trends in Cell Biology 11:442-445, 2001.*
116. Huang Z-y, Baldwin RL, Hedrick NM, Gutmann DH: *Astrocyte-specific expression of CDK4 is not sufficient for tumor formation, but cooperates with p53 heterozygosity to provide a growth advantage for astrocytes in vivo. Oncogene 21:1325-1334, 2002.*
117. Gutmann DH, Huang Z-y, Hedrick NM, Ding H, Guha A, Watson MA: *Mouse glioma gene expression profiling identifies novel human glioma-associated genes. Annals of Neurology 51:393-405, 2002.*
118. Uhlmann EJ, Apicelli AJ, Baldwin RL, Burke SP, Bajenaru ML, Onda H, Kwiatkowski D, Gutmann DH: *Heterozygosity for the tuberous sclerosis complex (TSC) gene products results in increased astrocyte numbers and decreased p27-Kip1 expression in TSC2+/- cells. Oncogene 21:4050-4059, 2002.*

119. Chan CC, Koch CA, Kaiser-Kupfer MI, Parry DM, Gutmann DH, Zhuang Z, Vortmeyer AO: Loss of heterozygosity for the NF2 gene in retinal and optic nerve lesions of patients with neurofibromatosis 2. *J. Pathol.* 198:14-20, 2002.
120. Ferner RE, Gutmann DH, Coffin C, Grimer R, Guha A, Judson I, Sandison A, Smith M, Viskochil D. International Consensus Statement on Malignant Peripheral Nerve Sheath Tumors in Neurofibromatosis 1. *Cancer Res* 62:1573-1577, 2002.
121. Singh PK, Gutmann DH, Fuller CE, Newsham IF, Perry A: Differential involvement of Protein 4.1 family members, DAL-1 and NF2 in intracranial and intraspinal ependymomas. *Modern Pathol.* 15:526-31, 2002.
122. Friedman JM, Arbiser J, Epstein JA, Gutmann DH, Huot SJ, Lin A, McManus B, Korf BR: Cardiovascular disease in neurofibromatosis 1: Report of the NF1 cardiovascular task force. *Genetics in Medicine* 4:105-11, 2002.
123. Packer RJ, Gutmann DH, Rubenstein A, Viskochil D, Zimmerman RA, Vezina G, Small J, Korf B: Plexiform neurofibromas in NF1: Towards biologic-based therapy. *Neurology* 58:1461-1470, 2002.
124. Gutmann DH, Hedrick NM, Li J, Nagarajan R, Perry A, Watson MA. Comparative gene expression profile analysis of neurofibromatosis 1 (NF1)-associated and sporadic pilocytic astrocytomas. *Cancer Res.* 62:2085-2091, 2002.
125. Kalamarides M, Niwa-Kawakita M, Leblois H, Abramowski V, Perricaudet M, Janin A, Thomas G, Gutmann DH, Giovannini M. Nf2 gene inactivation in arachnoidal cells is rate-limiting for meningioma development in the mouse. *Genes & Development* 16:1060-1065, 2002.
126. Baser ME, De Rienzo A, Altomare D, Balsara BR, Hedrick NM, Gutmann DH, Pitts LH, Jackler RK, Testa JR. Neurofibromatosis 2 and malignant mesothelioma. *Neurology* 59:290-291, 2002.
127. Bajenaru ML, Zhu Y, Hedrick NM, Donahoe J, Parada LF, Gutmann DH. Astrocyte-Specific Inactivation of the Neurofibromatosis 1 gene (NF1) is insufficient for astrocytoma formation. *Molecular and Cellular Biology* 22:5100-5113, 2002.
128. Uhlmann EJ, Wong M, Baldwin RL, Bajenaru ML, Onda H, Kwiatkowski DJ, Yamada K, Gutmann DH. Astrocyte-specific TSC1 conditional knockout mice exhibit abnormal neuronal organization and seizures. *Annals of Neurology* 52:285-296, 2002.
129. Yu T, Robb VA, Singh V, Gutmann DH, Newsham IF. The 4.1/ezrin/radixin/moesin domain of the DAL-1/Protein 4.1B tumour suppressor interacts with 14-3-3 proteins. *Biochem. J.* 365:783-9, 2002.
130. Watson MA, Gutmann DH, Peterson K, Chicoine MR, Kleinschmidt-DeMasters BK, Brown HG, Perry A. Molecular characterization of human meningiomas by gene expression profiling using high density oligonucleotide microarrays. *Am. J. Pathol.* 161:665-72, 2002.
131. Perry A, Kunz SN, Fuller CE, Banerjee R, Marley EF, Liapis H, Watson MA, Gutmann DH. Differential NF1, p16, and EGFR patterns by interphase cytogenetics (FISH) in malignant peripheral nerve sheath tumor (MPNST) and morphologically similar spindle cell neoplasms. *J. Neuropathol Exp Neurol.* 61:702-9, 2002.

132. Woods SA, Marmor E, Feldkamp MM, Lau N, Apicelli AJ, Boss G, Gutmann DH, Guha A. Aberrant G-protein signaling in nervous system tumors. *J. Neurosurg.* 97:627-42, 2002.
133. Gutmann DH, Rasmussen SA, Wolkenstein P, MacCollin MM, Guha A, Inskip PD, North KN, Poyhonen M, Birch PH, Friedman JM. Gliomas presenting after age ten in individuals with neurofibromatosis type 1 (NF1). *Neurology* 59:759-761, 2002.
134. Gutmann DH, Giovannini M: Mouse models of NF1 and NF2. *Neoplasia* 4:279-90, 2002.
135. Sun CX, Robb VA, Gutmann DH: Protein 4.1 tumor suppressors: Getting a FERM grip on growth regulation. *J. Cell Sci.* 115:3991-4000, 2002.
136. Sun CX, Haipek C, Scoles DR, Pulst SM, Giovannini M, Komada M, Gutmann DH: Functional analysis of the relationship between the neurofibromatosis 2 (NF2) tumor suppressor and its binding partner, hepatocyte growth factor-regulated tyrosine kinase substrate (HRS/HGS). *Human Molecular Genetics* 11:3167-3178, 2002.
137. Scoles DR, Nguyen VD, Qin Y, Sub C-X, Morrison H, Gutmann DH, Pulst S-M: Neurofibromatosis 2 (NF2) tumor suppressor schwannomin and its signaling protein HRS regulate STAT signaling. *Human Molecular Genetics* 11:3179-3189, 2002.
138. Lau N, Uhlmann EJ, von Lintig FC, Nagy A, Boss GR, Gutmann DH, Guha A: Rap1 activity is elevated in malignant astrocytomas independent of tuberous sclerosis complex-2 gene expression. *Int. J. Oncol.* 22: 195-200, 2003.
139. Huang Z-y, Wu Y, Hedrick N, Gutmann DH: T-cadherin-mediated cell growth regulation involves G<sub>2</sub> phase arrest and requires p21<sup>CIP1/WAF1</sup> expression. *Mol Cell Biol.* 23:566-578, 2003.
140. Apicelli AJ, Uhlmann EJ, Baldwin RL, Ding H, Nagy A, Guha A, Gutmann DH: The role of Rap1 GTPase in astrocyte growth regulation. *GLIA* 42:225-234, 2003.
141. Ding H, Shannon P, Lau N, Wu X, Roncari L, Baldwin RL, Takebayashi H, Nagy A, Gutmann DH, Guha A: Oligodendrogliomas result from the expression of an activated mutant epidermal growth factor receptor in a RAS transgenic mouse astrocytoma model. *Cancer Res* 63:1106-1113, 2003.
142. King A, Listernick R, Charrow J, Piersall L, Gutmann DH: Optic pathway gliomas in neurofibromatosis type 1: The effect of presenting symptoms on outcome. *American Journal of Medical Genetics* 122:95-99, 2003.
143. Gutmann DH, Baker SJ, Giovannini M, Garbow J, Weiss W: Mouse Models of Human Cancers Consortium (MMHCC) symposium on nervous system tumors. *Cancer Res.* 63:3001-3004, 2003.
144. Robb VA, Li W, Gascard P, Perry A, N Mohandas, Gutmann DH: Identification of a third Protein 4.1 tumor suppressor, Protein 4.1R, in meningioma pathogenesis. *Neurobiol Dis.* 13:191-202, 2003.
145. Huang Z-y, Wu Y, Burke SP, Gutmann DH: The 43-kDa growth associated protein functions as a negative growth regulator in glioma. *Cancer Res.* 63:2933-2939, 2003.

146. Wong M, Ess KC, Uhlmann EJ, Jansen LA, Li W, Crino PB, Mennerick S, Yamada KA, Gutmann DH: *Impaired astrocyte glutamate transport in a mouse epilepsy model of tuberous sclerosis. Annals of Neurology 54:251-256, 2003.*
147. Gutmann DH, Winkeler E, Kabbarah O, Hedrick N, Dudley S, Goodfellow PJ, Liskay RM: *Mlh1 deficiency accelerates myeloid leukemogenesis in neurofibromatosis 1 (Nf1) heterozygous mice. Oncogene 22:4581-4585, 2003.*
148. Maldonado M, Newman D, Kolson DL, Chen W, Gutmann DH, Crino PB: *Specific expression of ICAM-1, TNF- $\alpha$ , NF B and MAPK in human tuberous sclerosis complex (TSC)-associated tubers and in mice with conditional Tsc1 inactivation. Neurobiol. Dis. 14:279-290, 2003.*
149. Angel P, Tabancay AP, Gau CL, Machado IMP, Uhlmann EJ, Gutmann DH, Guo L, Tamanoi F: *Identification of dominant negative mutants of Rheb GTPase and their use to implicate the involvement of human Rheb in the activation of p70S6K. J. Biol. Chem. 278:39921-30, 2003.*
150. Gutmann DH, James CD, Poyhonen M, Louis DN, Ferner R, Guha A, Hariharan S, Viskochil DV, Perry A: *Molecular analysis of astrocytomas presenting after age 10 in individuals with NF1. Neurology 61:1397-1400, 2003.*
151. Dasgupta B, Dugan LL, Gutmann DH: *The neurofibromatosis 1 gene product neurofibromin regulates Pituitary Adenylate Cyclase-Activating Polypeptide-mediated signaling in astrocytes. J. Neurosci. 23:8949-54, 2003.*
152. Oh M-K, Scoles DR, Haipek C, Strand AD, Gutmann DH, Olson JM, Pulst S-M: *Genetic heterogeneity of stably transfected cell lines revealed by expression profiling with oligonucleotide microarrays. J. Cellular Biochem. 90:1068-78, 2003.*
153. Bajenaru ML, Hernandez MR, Perry A, Zhu Y, Parada LF, Garbow JR, Gutmann DH: *Optic nerve glioma in mice requires astrocyte Nf1 gene inactivation and Nf1 brain heterozygosity. Cancer Res. 63: 8573-8577, 2003.*
154. Surace EI, Haipek CA, Gutmann DH: *The effect of merlin phosphorylation on neurofibromatosis 2 (NF2) gene function. Oncogene 23: 580-7, 2004.*
155. Ess KC, Uhlmann EJ, Li W, Li H, DeClue JE, Crino PB, Gutmann DH: *Expression profiling in tuberous sclerosis complex (TSC) knockout mouse astrocytes to characterize human TSC brain pathology. GLIA 46: 28-40, 2004.*
156. Spritz RA, Itin PH, Gutmann DH: *Piebaldism and Neurofibromatosis Type 1: Horses of Very Different Colors. J. Invest. Dermatol. 122:xxxiv-xxxv, 2004.*
157. Su W, Gutmann DH, Perry A, Abounader R, Laterra J, Sherman LS: *A CD44-independent hepatocyte growth factor/c-Met autocrine loop promotes malignant peripheral nerve sheath tumor cell invasion. GLIA 45: 297-306, 2004.*
158. Chen Y, Gutmann DH, Haipek CA, Martinsen BJ, Bronner-Fraser M, Krull CE: *Characterization of chicken Nf2/merlin indicates regulatory roles in cell proliferation and migration. Developmental Dynamics 229:541-54, 2004.*
159. Robb, VA, Li W, Gutmann DH: *Disruption of 14-3-3 binding does not impair Protein 4.1B growth suppression. Oncogene 23:3589-96, 2004.*

160. Uhlmann EJ, Li W, Scheidenhelm D, Gau C-L, Tamanoi F, Gutmann DH: Loss of tuberous sclerosis complex 1 (*Tsc1*) expression results in increased Rheb/S6K pathway signaling important for astrocyte cell size regulation. *GLIA* 47:180-8, 2004.
161. Fedi M, Mitchell LA, Renate M Kalnins RM, Gutmann DH, Perry A, Newton M, Brodtmann A, Berkovic SF: Glioneuronal Tumors in Neurofibromatosis Type 1: MRI-Pathological Study. *J Clin. Neurosci.* 11:745-7, 2004.
162. Rong R, Surace EI, Haipek CA, Gutmann DH, Ye K: Serine 518 phosphorylation modulates merlin intramolecular association and binding to critical effectors important for NF2 growth suppression. *Oncogene* 23:8447-54, 2004.
163. Surace EI, Lusic E, Murakami Y, Scheithauer BW, Perry A, Gutmann DH: Loss of tumor suppressor in lung cancer-1 (*TSLC1*) expression in meningioma correlates with increased malignancy grade and reduced patient survival. *J. Neuropathol. Exp. Neurol.* 63:1015-1027, 2004.
164. Surace EI, Lusic E, Haipek CA, Gutmann DH: Functional significance of S6K overexpression in meningioma progression. *Annals of Neurology* 56:295-98, 2004.
165. Watson MA, Perry A, Tihan T, Prayson RA, Guha A, Bridge J, Ferner R, Gutmann DH: Gene expression profiling reveals unique molecular subtypes of neurofibromatosis type 1-associated and sporadic malignant peripheral nerve sheath tumors. *Brain Pathol.* 14: 297-303, 2004.
166. Listernick R, Ferner RE, Piersall LS, Sharif S, DH Gutmann DH, Charrow J: Late-onset optic pathway tumors in children with neurofibromatosis 1 (NF1). *Neurology* 63:1944-1946, 2004.
167. Singh V, Miranda TB, Jiang W, Frankel A, Roemer ME, Robb VA, Gutmann DH, Herschman HR, Clarke S, Newsham IF: The DAL-1/4.1B tumor suppressor interacts with protein arginine N-methyltransferase 3 (PRMT3) and inhibits its ability to methylate substrates in vitro and in vivo. *Oncogene* 23:7761-71, 2004.
168. Fraser MM, Zhu X, Kwon CH, Uhlmann EJ, Gutmann DH, Baker SJ: Pten loss causes hypertrophy and increased proliferation of astrocytes in vivo. *Cancer Research* 64:7773-9, 2004.
169. Rong R, Gutmann DH, Ye K: The neurofibromatosis 2 tumor suppressor, merlin, inhibits PI 3-kinase through binding to PIKE-L. *Proceedings of the National Academy of Sciences USA* 101:18200-18205, 2004.
170. Bajenaru ML, Garbow JR, Perry A, Hernandez MR, Gutmann DH: Natural history of neurofibromatosis 1-associated optic nerve glioma formation in mice. *Annals of Neurology* 57:119-27, 2005.
171. Dasgupta B, Li W, Perry A, Gutmann DH: Glioma formation in neurofibromatosis 1 reflects preferential activation of K-RAS in astrocytes. *Cancer Research* 65:236-45, 2005.
172. Robb VA, Gerber MA, Hart-Mahon EK, Gutmann DH: Membrane localization of the U2 domain of Protein 4.1B is necessary and sufficient for meningioma growth suppression. *Oncogene* 24:1946-57, 2005.

173. Kabbarah O, Sotelo AK, Mallon AN, Winkeler EL, Fan M-Y, Pfeiffer JD, Shibata D, Gutmann DH, Goodfellow PJ: Diethylstilbesterol effects and lymphomagenesis in Mlh1-deficient mice. *Int. J. Cancer* 115:666-669, 2005.
174. Perry A, Kurtkaya-Yapicier O, Scheithauer BW, Robinson S, Prayson RA, Kleinschmidt-DeMasters BK, Stemmer-Rachamimov AO, Gutmann DH: Insights into meningioangiomas with and without meningioma: A clinicopathologic and genetic series of 24 cases with a review of the literature. *Brain Pathology* 15:55-65, 2005.
175. Ess KC, Kamp CA, Tu BP, Gutmann DH: Developmental origin of subependymal giant cell astrocytoma in tuberous sclerosis complex. *Neurology* 64:1446-1449, 2005.
176. Scheidenhelm DK, Cresswell J, Haipek CA, Fleming TP, Mercer RW, Gutmann DH: Akt-dependent cell size regulation by the adhesion molecule on glia (AMOG) occurs independently of phosphatidylinositol 3-kinase and Rheb signaling. *Mol. Cell. Biol.* 25:3151-62, 2005.
177. Rajaram V, Gutmann DH, Prasad SK, Mansur D, Conboy JG, Perry A: Altered Protein 4.1 family member expression in ependymomas: A study of 84 cases. *Modern Pathology* 18:991-7, 2005.
178. Dasgupta B, Yi Y, Chen DY, Weber JD, Gutmann DH: Proteomic analysis reveals hyperactivation of the mTOR pathway in NF1-associated human and mouse brain tumors. *Cancer Research* 65:2755-60, 2005.
179. Perry A, Lusic E, Gutmann DH: Meningothelial hyperplasia: A detailed clinicopathologic, immunohistochemical, and genetic study of 11 cases. *Brain Pathology* 15:109-15, 2005.
180. Rubin JB, Gutmann DH: Neurofibromatosis 1: A model for nervous system tumour formation? *Nature Reviews Cancer* 5:557-564, 2005.
181. Yang G, Khalaf W, van de Locht L, Jansen JH, Gao M, Thompson MA, van der Reijden BA, Gutmann DH, Delwel R, Clapp DW, Hiebert SW: Transcriptional repression of the Neurofibromatosis-1 tumor suppressor by the t(8;21) fusion protein. *Mol. Cell. Biol.* 25:5869-79, 2005.
182. Dasgupta B, Gutmann DH: Neurofibromin regulates neural stem cell proliferation, survival, and astroglial differentiation in vitro and in vivo. *J. Neurosci.* 25:5584-94, 2005.
183. Kuns R, Kissil JL, Newsham IF, Jacks T, Gutmann DH, Sherman LS: Protein 4.1B expression is induced in mammary epithelial cells during pregnancy and regulates their proliferation. *Oncogene* 24:6502-15., 2005.
184. Lusic EA, Watson MA, Chicoine MR, Lyman M, Roerig P, Reifenberger G, Gutmann DH, Perry A: Integrative Genomic Analysis Identifies NDRG2 as a Candidate Tumor Suppressor Gene Frequently Inactivated in Clinically-Aggressive Meningioma. *Cancer Res.* 65:7121-6, 2005.
185. Shannon P, Sabha N, Lau N, Kamnasaran D, Gutmann DH, Guha A: Pathological and molecular progression of astrocytomas in a GFAP:12V-Ha-RAS mouse astrocytoma model. *Am. J. Pathol.* 167:859-67, 2005.
186. Yamada D, Kikuchi S, Williams YN, Sakurai-Yageta M, Masuda M, Maruyama T, Tomita K, Gutmann DH, Kakizoe T, Kitamura T, Kanai Y, Murakami Y: Promoter

*Hypermethylation of the DAL-1/4.1B Gene is Correlated with Poor Prognosis in Renal Clear Cell Carcinoma. Int. J. Cancer 118:916-23, 2005.*

187. Jansen LA, Uhlmann EJ, Crino PB, Gutmann DH, Wong M: *Epileptogenesis and reduced inward rectifier potassium current in tuberous sclerosis complex-1 deficient astrocytes. Epilepsia 46:1871-80, 2005.*
188. Sharma MK, Zehnbaauer BA, Watson MA, Gutmann DH: *RAS pathway activation and an oncogenic RAS mutation in sporadic pilocytic astrocytoma. Neurology 65:1335-6, 2005.*
189. Scoles DR, Qin Y, Nguyen V, Gutmann DH, Pulst SM: *HRS inhibits EGF receptor signaling in the RT4 rat schwannoma cell line. Biochemical and Biophysical Research Communications 335:385-392, 2005.*
190. Dasgupta B, Yi Y, Hegedus B, Weber JD, Gutmann DH: *Cerebrospinal fluid proteomic analysis reveals dysregulation of methionine aminopeptidase-2 expression in human and mouse neurofibromatosis 1-associated glioma. Cancer Res. 65:9843-50, 2005.*
191. Mawrin C, Schulz S, Hellwig-Pattyk A, Kirches E, Roessner A, Lendeckel U, Firsching R, Vorwerk CK, Keilhoff G, Dietzmann K, Kluwe L, Lindberg G, Gutmann DH, Scheithauer B, Perry A: *Expression and Function of Somatostatin Receptors in Peripheral Nerve Sheath Tumors. J Neuropath Exp Neurol. 64: 1080-1088, 2005.*
192. Zhu Y, Harada T, Liu L, Lush ME, Guignard F, Harada C, Burns DK, Bajenaru ML, Gutmann DH, Parada LF: *Inactivation of NF1 in CNS causes increased glial progenitor proliferation and optic glioma formation. Development 132:5577-5588, 2005.*
193. Mangoura D, Sun Y, Li C, Singh D, Gutmann DH, Flores A, Ahmed M, Villianatos G: *Phosphorylation of neurofibromin by PKC is a possible molecular switch in EGF receptor signalling in neural cells. Oncogene 25:735-45, 2006.*
194. Sharma MK, Watson MA, Lyman M, Perry A, Aldape KD, Deak F, Gutmann DH: *Matrilin-2 expression distinguishes clinically relevant subsets of pilocytic astrocytoma. Neurology 66:127-130, 2006.*
195. Gutmann DH, Maher EA, Van Dyke T: *Mouse Models of Human Cancers Consortium Workshop of Nervous System Tumors. Cancer Res. 66:10-13, 2006.*
196. Miller SJ, Rangwala F, Williams J, Ackerman P, Kong S, Jegga A, Aronow B, Frahm S, Kluwe L, Mautner V, Upadhyaya M, Muir D, Wallace M, Hagen J, Quelle DE, Watson MA, Perry A, Gutmann DH, Ratner N: *Large-scale molecular comparison of human Schwann cells to malignant peripheral nerve sheath tumor cells and tissues. Cancer Res. 66:2584-91, 2006.*
197. Chadee DN, Hung G, Andalibi A, Lim DJ, Luo Z, Gutmann DH, Kyriakis JM: *MLK3 regulates B-Raf through a mechanism that involves maintenance of the B-Raf/Raf-1 complex and inhibition by the NF2 tumor suppressor protein. Proc. Natl. Acad. Sci. USA 103:4463-8, 2006.*
198. Gutmann DH, Hunter-Schaedle K, Shannon KM: *Harnessing preclinical mouse models to inform human clinical cancer trials. J. Clin. Invest. 116:847-852, 2006.*
199. Gerber MA, Bahr SM, Gutmann DH: *Protein 4.1B/DAL-1 functions as a growth suppressor in meningioma cells by activating Rac1-dependent JNK signaling. Cancer Res. 66:5295-303, 2006.*

200. Read TA, Hegedus B, Wechsler-Reya R, Gutmann DH: *The Neurobiology of Neuro-Oncology*. *Ann. Neurol.* 60:3-11, 2006.
201. Wei Q, Clarke L, Scheidenhelm DK, Qian B, Tang A, Sabha N, Karim Z, Bock NA, Reti R, Swoboda R, Purev E, Bajenaru ML, Shannon P, Herlyn D, Kaplan D, Henkelman M, Gutmann DH, Guha A: *High-grade glioma formation results from postnatal Pten loss or mutant epidermal growth factor receptor expression in a transgenic mouse glioma model*. *Cancer Research* 66:7429-37, 2006.
202. Leonard JR, Perry A, Rubin JB, King AA, Chicoine MR, Gutmann DH: *The role of surgical biopsy in the diagnosis of glioma in individuals with neurofibromatosis-1*. *Neurology* 67:1509-1512, 2006.
203. Surace EI, Strickland A, Gutmann DH, Naughton CK: *Tslc1 (nectin-like molecule-2) is essential for spermatozoa motility and male fertility*. *J. Androl.* 27:816-825, 2006.
204. Houshmandi SS, Surace EI, Zhang H, Fuller GN, Gutmann DH: *Tumor suppressor in lung cancer-1 (TSLC1) functions as a glioma tumor suppressor*. *Neurology* 67:1863-66, 2006.
205. Spinner RJ, Scheithauer BW, Perry A, Amrami KK, Emmett R, Gutmann DH: *Co-localized cellular schwannoma and plexiform neurofibroma in the absence of neurofibromatosis*. *J. Neurosurgery* (in press).
206. Sharma MK, Mansur DB, Reifenberger G, Perry A, Leonard JR, Aldape KD, Albin MG, Emmett RJ, Loeser S, Watson MA, Nagarajan R, Gutmann DH: *Distinct genetic signatures among pilocytic astrocytomas relate to their brain region origin*. *Cancer Res.* 67:890-900, 2007.
207. Pelletier CL, Maggi LB, Scheidenhelm DK, Gutmann DH, Weber JD: *Tuberous Sclerosis Complex-1 sets the rate of ribosome export and protein synthesis through nucleophosmin translation*. *Cancer Res.* 67:1609-17, 2007.
208. Su W, Xing R, Guha A, Gutmann DH, Sherman LS: *Mice with GFAP-targeted loss of neurofibromin demonstrate increased axonal MET expression with aging*. *GLIA* 55:723-33, 2007.
209. Listernick R, Ferner RE, Liu GT, Gutmann DH: *Optic pathway gliomas in neurofibromatosis-1: Controversies and recommendations*. *Annals of Neurology* 61:189-98, 2007.
210. Sandsmark DK, Pelletier C, Weber JD, Gutmann DH: *Mammalian target of rapamycin: Master regulation of cell growth in the nervous system*. *Histology and Histopathology* 22:895-903, 2007.
211. Erbayat-Altay E, Zeng L-H, Xu L, Gutmann DH, Wong M: *The natural history and treatment of epilepsy in a murine model of tuberous sclerosis*. *Epilepsia* (in press).
212. Banerjee D, Hegedus B, Gutmann DH, Garbow JR: *Detection and measurement of neurofibromatosis-1 mouse optic glioma in vivo*. *Neuroimage* 35:1434-7, 2007.
213. Dagainakatte GC, Gutmann DH: *Neurofibromatosis-1 (Nf1) heterozygous brain microglia elaborate paracrine factors that promote Nf1-deficient astrocyte and glioma growth*. *Human Molecular Genetics* 16:1098-1112, 2007.

214. Sandsmark DK, Zhang H, Hegedus B, Pelletier CL, Weber JD, Gutmann DH: Nucleophosmin mediates mammalian target of rapamycin-dependent actin cytoskeleton dynamics and proliferation in neurofibromin-deficient astrocytes. *Cancer Research* 67:4790-9, 2007.
215. Leonard JR, Ferner RE, Thomas N, Gutmann DH: Cervical cord compression from plexiform neurofibromas in neurofibromatosis-1. *Journal of Neurology, Neurosurgery, and Psychiatry* (in press).
216. Zheng L-H, Ouyang Y, Gazit V, Cirrito JR, Jansen LA, Ess KC, Yamada KA, Wozniak DF, Holtzman DM, Gutmann DH, Wong M: Abnormal glutamate homeostasis and impaired synaptic plasticity and learning in a mouse model of tuberous sclerosis complex. *Neurobiol. Dis.* (in press).
217. Warrington NM, Woerner BM, Daginakatte GC, Dasgupta B, Perry A, Gutmann DH, Rubin JB: Spatiotemporal differences in CXCL12 expression and cyclic AMP underlie the unique pattern of optic glioma growth in neurofibromatosis type 1. *Cancer Res.* (in press).
218. Maurel P, Einheber S, Galinska J, Thaker P, Lam I, Rubin MB, Scherer SS, Murakami Y, Gutmann DH, Salzer JL: Nectin-like proteins mediate axon-Schwann cell interactions along the internode and are essential for myelination. *J. Cell Biol.* (in press).
219. Hegedus B, Dasgupta B, Shin JE, Emmett RJ, Hart-Mahon EK, Elghazi L, Bernal-Mizrachi E, Gutmann DH: Neurofibromatosis-1 regulates neuronal and glial cell differentiation from neuroglial progenitors in vivo by both cAMP- and Ras-dependent mechanisms. *Cell Stem Cell* (in press).
220. Tang X, Jang S-W, Wang X, Liu Z, Bahr SM, Sun S-Y, Brat D, Gutmann DH, Ye K: Akt phosphorylation regulates merlin tumor suppressor activity by mediating merlin ubiquitination and degradation. *Nature Cell Biology* (in press).

Invited Publications:

Reviews:

1. Gutmann DH, Collins FS: The neurofibromatosis type 1 gene product and its protein product, neurofibromin. *Neuron* 10:335-343, 1993.
2. Gutmann DH, Collins FS: The neurofibromatosis type 1 (NF1) gene: Beyond positional cloning. *Arch. Neurol* 50:1185-1193, 1993.
3. Gutmann DH: The neurofibromatoses: New insights into old problems. *Current Opinions in Neurology* 7:166-171, 1996.
4. Gutmann DH: Cell Motility and the Neurofibromatosis 2 tumor suppressor gene. *Neurology Network Commentary* 1: 191-196, 1997.
5. Gutmann DH: Recent insights into neurofibromatosis 1: clear genetics progress. *Archives of Neurology* 55:778, 1998.
6. Gutmann DH: The TSC2 gene product, tuberlin, functions as a Rab5 GTPase activating protein (GAP) in modulating endocytosis. *Neurology Network Commentary* 1:291-295, 1998.

7. Gutmann DH: *New insights into the molecular pathogenesis of Huntington Disease: expanding on a theme. Neurology Network Commentary 2:111-117, 1998.*
8. Gutmann DH: *The diagnosis and management of neurofibromatosis 1. The Neurologist 4:313-326, 1998.*
9. Leventer RJ, Gutmann DH: *Recent insights into the molecular genetics of human brain development. Neurology Network Commentary 3:127-132, 1999.*
10. Gutmann DH: *Learning disabilities in neurofibromatosis 1: sizing up the brain. Arch Neurol 56: 1322-1323, 1999.*
11. King A, Gutmann DH: *The question of familial meningiomas and schwannomas: NF2B or not to be? Neurology 54:4-5, 2000.*
12. Ding H, Nagy A, Gutmann DH, Guha A: *Astrocytoma models. Neurosurgical Focus 8:1-7, 2000.*
13. Devere T, Gutmann DH: *Channeling the episodic ataxias. Neurology Network Commentary www.lancetneuronet.com 2001;4(2).*
14. Gutmann DH: *Neurofibromin in the brain: Working RAS backwards. J. Child Neurol. 17:592-601, 2002.*
15. Baser ME, Evans DGR, Gutmann DH: *Neurofibromatosis 2. Current Opinion in Neurology. 16:27-33, 2003.*
16. Dasgupta B, Gutmann DH: *Neurofibromatosis 1: closing the GAP between mice and men. Current Opinion in Genetics and Development. 13:20-27, 2003.*
17. Arun D, Gutmann DH: *Neurofibromatosis 1. Current Opinion in Neurology. 17:101-105, 2004.*
18. Perry A, Gutmann DH, Reifenberger G. *Molecular pathogenesis of meningiomas. J. Neuro-oncol. 70:183-202, 2004.*
19. Scheidenhelm DK, Gutmann DH: *Mouse models of tuberous sclerosis complex. J. Child Neurol. 19:726-33, 2004.*
20. Maria B, Deidrick KM, Roach ES, Gutmann DH: *Tuberous sclerosis complex: pathogenesis, diagnosis, strategies, therapies, and future research directions. J. Child Neurol. 19:632-42, 2004.*
21. Lusi E, Gutmann DH: *Meningioma: An Update. Current Opinion in Neurology. 17:687-692, 2004.*
22. Ward BA, Gutmann DH: *Neurofibromatosis 1 - From Lab Bench to Clinic. Pediatr. Neurol. 32:211-8, 2005.*
23. Gilbertson RJ, Gutmann DH: *Tumorigenesis in the brain: Location, location, location. Cancer Res. 67:5579-82, 2007.*
24. Houshmandi SS, Gutmann DH: *All in the family: Using inherited cancer syndromes to understand de-regulated cell signaling in brain tumors. J. Cell. Biochem. (in press).*

Book chapters:

1. Schook LB, Gutmann DH, Niederhuber JE: *Characterization of bone marrow derived macrophages as Ia-bearing accessory cells. Advances in Experimental Medicine and Biology* 149:429-433, 1982.
2. Schook LB, Gutmann DH, Marlin LE, Niederhuber JE: *Expression of Ia antigens and Ir gene function during differentiation of bone marrow derived macrophages. In Ir Genes: Past Present and Future (Pierce CW, et al., editors) Humana Press: New Jersey; pp. 91-101, 1983.*
3. Gutmann DH, Allen PM, Niederhuber JE: *Separation of immune response genes for LDH-B and MOPC-173: Implications for a new model of I-region gene organization. In Ir Genes: Past Present and Future (Pierce CW, et al., editors) Humana Press: New Jersey; pg 91-95, 1983.*
4. Gutmann DH, Fischbeck KH, Sladky JT: *The congenital muscle diseases. In Fetal and Neonatal Physiology (Polin R and Fox W, editors). WB Saunders: Philadelphia, pp 1675-1679, 1991.*
5. Gutmann DH, Collins FS: *von Recklinghausen neurofibromatosis. In Metabolic Basis of Inherited Diseases. (Scriver CR, Beaudet AL, Sly WS, Valle D, editors). 7th edition. McGraw-Hill: New York. pp. 677-696, 1994.*
6. Gutmann DH, Cole JL, Collins FS: *Expression of the neurofibromatosis type 1 (NF1) gene during mouse embryonic development. Progress in Brain Research* 105:327-335, 1995.
7. Riccardi VM, Gutmann DH: *The clinical and molecular genetics of neurofibromatosis 1 and neurofibromatosis 2. In The Molecular and Genetic Basis of Neurological Disease (Rosenberg RN, editor), 2<sup>nd</sup> edition, Chapter 40:693-712, 1997.*
8. Gutmann DH, Pulst SM: *The Phakomatoses. In: Neurogenetics. F.A. Davis. Chapter 9 1997.*
9. Gutmann DH: *Molecular biology of neurofibromatosis 1. In: Neurofibromatosis type 1 in childhood. (North K, editor). MacKeith Press: London , pp. 5-15, 1997.*
10. Gutmann DH, Collins FS: *von Recklinghausen neurofibromatosis. In: Metabolic Basis of Inherited Diseases. (Scriber CR, Beaudet AL, Sly WS, Valle D, editors). 7th Edition CD-ROM update. McGraw-Hill: New York, 1997.*
11. Gutmann DH, Collins FS: *Neurofibromatosis 1. In "The Genetic Basis of Human Cancer" (Vogelstein B and Kinzler KW, editors). McGraw-Hill: New York. Pages 423-442, 1997.*
12. Hsu CY, Lin TN, Gutmann DH: *Molecular biology and genetics in neurological disorders. In: Textbook of Neurology. (Marc Fisher and Julien Bogousslavsky, eds.). Butterworth-Heineman, pp. 127-133, 1998.*
13. Gutmann DH: *Neurofibromatosis 1: progress and future directions. In: Neurofibromatosis Type 1: from Genotype to Phenotype. Upadhyaya M. and Cooper DN (editors) Bios Scientific: Oxford, pp. 215-224, 1998.*

14. Gutmann DH: *Parallels between tuberous sclerosis complex and neurofibromatosis 1*. In: *Neurocutaneous Disorders: Seminars in Pediatric Neurology* (R.S. Kandt, editor) 5:276-286, 1998.
15. Gutmann DH, Riccardi VM: *Neurofibromatosis*. In: *Prognosis of Neurological Disorders, 2<sup>nd</sup> edition* (RW Evans, DS Baskin, FM Yatsu, editors). Chapter 31, 1999.
16. Gutmann DH, Gurney JG: *Other malignancies*. In: *Neurofibromatosis 3<sup>rd</sup> edition* (Friedman JM, et al. editors). Johns Hopkins Press. Chapter 10, 1999.
17. Gutmann DH: *Nervous system abnormalities*. In: *Neurofibromatosis 3<sup>rd</sup> edition* (Friedman JM, et al. editors). Johns Hopkins Press. Chapter 8, 1999.
18. Listerneck R, Gutmann DH: *Optic pathway tumors*. In: *Neurofibromatosis 3<sup>rd</sup> edition* (Friedman JM, et al. editors). Johns Hopkins Press. Chapter 9, 1999.
19. Pulst S, Gutmann DH: *Phakomatoses*. In *Neurogenetics: Contemporary Neurology Series, Volume 57, Chapter 9, pp. 163-189, 1999*.
20. MacCollin MM, Gutmann DH: *Brain tumors associated with neurofibromatosis*. In *Brain Tumors: An Encyclopedic Approach* (Kaye and Laws, editors), Churchill-Livingstone, London 2001.
21. Perry A, Leonard JR, Roth KA and Gutmann DH: *Tumor Genetics*. In *Textbook of Neurological Surgery*. (Batjer and Loftus, editors). Lippincott-Raven Publishers, Philadelphia 2002.
22. Colman H, Gutmann DH: *Neurofibromatosis 1 and 2*. In *Current Therapy in Neurological Diseases, Volume 6* (Johnson RT, Griffin JW, McArthur JC, eds), pages 110-113, 2001.
23. Goldstein J, Gutmann DH: *Neurofibromatosis 1*. In *Neurocutaneous Syndromes*, (Roach ES, Miller VS, eds) Cambridge University Press; Cambridge. Pages 42-49, 2003.
24. Gutmann DH, Collins FS: *von Recklinghausen neurofibromatosis*. In: *Metabolic Basis of Inherited Diseases*. (Scriber CR, Beaudet AL, Sly WS, Valle D, editors). 8th Edition. McGraw-Hill: New York. Pages 877-896, 2001.
25. Ess KC, Gutmann DH: *Neurofibromatoses*. In: *Manual of Neurological Practice*. (R. Evans, editor). WB Saunders: Orlando. Pages 896-898, 2003.
26. Ess KC, Gutmann DH: *Tuberous sclerosis complex and other neurocutaneous disorders*. In: *Manual of Neurological Practice*. (R. Evans, editor). WB Saunders: Orlando. Pages 899-902, 2003.
27. Lynch TM, Gutmann DH: *Neurofibromatosis 1*. In: *Neurologic Clinics of North America*. 20:841-865, 2002.
28. McCullough LA, Gutmann DH: *GTPase activating proteins: Neurofibromatosis 1*. In: *Molecular Basis of Inborn Errors of Development*. Oxford University Press: San Francisco 2003.
29. Gurnett CA, Gutmann DH: *Neuro-oncology: The Neurofibromatoses*. In: *The Molecular and Genetic Basis of Neurological and Psychiatric Disease*. 3<sup>rd</sup> Edition (Rosenbery RN, et al., editors). Butterworth & Heinemann: MA. Pages 389-398, 2003.

30. *Esper GJ, Gutmann DH: Neurofibromatosis 1 and 2. In: Principles of Molecular Medicine. 2<sup>nd</sup> edition. (S. Strittmatter, et al., editors) Humana Press: NJ, 2004.*
31. *Gutmann DH, Perry A, Rangwala R, Sherman LS: Mouse models of Peripheral Nervous System Tumors. (E. Holland, editor). In: Mouse Models of Human Cancer. John Wiley and Sons: New Jersey pp189-198, 2004.*
32. *Gutmann DH, Wetmore C, O'Neill BP: The Phakomatoses. In: Principles of Neuro-Oncology (D. Schiff, editor) McGraw Hill: Philadelphia pp 233-256, 2005.*
33. *Malhotra A, Gutmann DH: Neurofibromatosis 1. In: Neurogenetics: Scientific and Clinical Advances. (D. Lynch, editor). Marcel Dekker, Inc.: New York. 2005.*
34. *Gutmann DH, Perry A: Neurofibromatosis 1. In: Russell & Rubinstein Pathology of Tumors of the Nervous System, 7<sup>th</sup> Edition, Hodder Arnold Press: London, Pages 903-916, 2006.*
35. *Leeman B, Gutmann DH: Neurofibromatosis 1. In: Current Pediatric Therapy. 18<sup>th</sup> Edition. (Burg, Ingelfinger, Polin, and Gershon, editors). Elsevier Press: Philadelphia, Pages 396-399, 2006.*
36. *Piersall L, Gutmann DH: Neurofibromatosis 1. In: Neurobiology of Disease (S. Gilman, editor) Elsevier: New York Pages 413-424, 2006.*
37. *Daginakatte GC, Gutmann DH: NF1 and Neurofibromatosis 1. In: Molecular Basis of Inborn Errors of Development. Oxford University Press: San Francisco. 2<sup>nd</sup> edition (in press).*
38. *Bogitch R, Gutmann DH: Neuro-oncology: The Neurofibromatoses. In: The Molecular and Genetic Basis of Neurological and Psychiatric Disease. 4<sup>th</sup> Edition (Rosenberg RN, et al., editors). Butterworth & Heinemann: MA. (in press).*
39. *Jost S, Gutmann DH: Neurofibromatosis and Other Genetic Syndromes. In: Handbook of Clinical Neurology, Neuro-Oncology Volume. (Aminoff, Biller and Swaab, editors). Elsevier: New York (in press).*
40. *Zeng L-H, Gutmann DH, Wong M: Glial-mediated mechanisms of epileptogenesis in tuberous sclerosis. In: Encyclopedia of Basic Epilepsy Research. (PA Schwartzkroin, editor). Elsevier: Oxford (in press).*

Brochures:

1. *Piersall L, Gutmann DH, Ferner R: Living with neurofibromatosis type 1: A guide for adults. National Neurofibromatosis Foundation, 2002.*
2. *Piersall L, Gutmann DH: Let's Talk About Neurofibromatosis: A guide for teens. Washington University, 2006.*
3. *Piersall L, Gutmann DH: Understanding Your Child's Diagnosis of Neurofibromatosis. Washington University, 2007.*

- c. Who are the key staff in your NF clinic facility?  
Provide Name; Title; Degree/Qualifications; Role in Clinic.

Anne C. Albers, RN, PNP. Anne is a pediatric nurse practitioner with over 20 years of experience caring for children. Anne serves as the Washington University NF Clinic Coordinator.

The list of our consultants can be found in section 3.

- d. Who within this core staff currently coordinates NF patient services? Describe this individual's NF clinic related duties.

Anne Albers functions as our clinic coordinator. She is primarily responsible for providing timely follow-up, handling phone calls, interacting with our consultants, and serving as a liaison to the NF community.

- e. Describe any areas of NF care in which your clinic has particular expertise (e.g. optic glioma, vestibular schwannoma, bone manifestations, learning disabilities etc.) and the clinic staff that provide this care.

The Washington University NF Clinic has extensive expertise in brain tumors in NF1 (optic glioma), malignant peripheral nerve sheath tumors, and plexiform neurofibromas.

### 3. PATIENT SCHEDULING and REFERRALS

- a. Provide the details of the 'typical' timeframe in which patients receive a response to a request for scheduling, are actually scheduled for an appointment, how patients are prioritized, etc.

Patients are prioritized based on the acuity of their medical needs. Patients are typically seen within 2-3 weeks; however, patients requiring more immediate care are seen urgently.

- b. Provide details of those specialists to whom (either within or outside our own clinic facility) your clinic refers NF patients for the following specialty care. These should be individuals familiar and experienced with consensus guidelines for care of individuals with NF (Please provide information for PEDIATRIC CARE referrals in the first table and ADULT CARE in the second table).

#### **PEDIATRIC CARE**

<b>SPECIALTY</b>	<b>DOCTOR</b>	<b>CLINIC ADDRESS</b>	<b>PHONE</b>	<b>EMAIL (if available)</b>
Genetics	Dr. Tyler Reimschisel	#1 Children's Place	314-454-6093	
Neurology	Dr. David Gutmann	#1 Children's Place	314-454-6120	
Orthopedics	Dr. Perry Schoenecker or Dr. Matthew Dobbs	#1 Children's Place	314-454-2500	

Developmental pediatrics/learning disabilities	Dr. James Keating	#1 Children's Place	314-454-2479	
Ophthalmology	Dr. Gregg Lueder	#1 Children's Place	314-454-6026	
Neurosurgery	Dr. Jeffrey Leonard	#1 Children's Place	314-454-4630	
Plastic surgery	Dr. Albert Woo	#1 Children's Place	314-362-7388	
Neuro-oncology	Dr. Joshua Rubin or Dr. Allison King	#1 Children's Place	314-454-6018	
Medical Oncology/Radiation Oncology	Dr. David Mansur	#1 Children's Place	314-454-7236	
Endocrinology	Dr. Abby Hollander	#1 Children's Place	314-454-6051	
Audiology/ENT	Dr. David Molter	#1 Children's Place	314-454-6162	
Radiology/Neuroradiology	Dr. Robert McKinstry	#1 Children's Place	314-454-2525	
General Surgery/Surgical Oncology	Dr. Patrick Dillon	#1 Children's Place	314-454-6022	
Dermatology	Dr. Susan Mallory	#1 Children's Place	314-454-2714	
Cardiovascular Disease	Dr. Achi Ludomirsky	#1 Children's Place	314-454-6095	
Oral and Maxillofacial Surgery	Dr. Mark Beehner	#1 Children's Place	314-362-4559	
Behavioral Issues	Dr. Jeffrey Titus	#1 Children's Place	314-454-6069	
Pain	Dr. Barry Jones	#1 Children's Place	314-454-6246	

## ADULT CARE

SPECIALTY	DOCTOR	CLINIC ADDRESS	PHONE	EMAIL (if available)
Genetics	Dr. Tyler Reimschisel	#1 Children's Place	314-454-2479	
Neurology	Dr. David Gutmann	#1 Children's Place	314-454-6120	
Orthopedics	Dr. Keith Bridwell	#1 Children's Place	314-747-2533	
Ophthalmology	Dr. William Hart	660 S. Euclid Ave	314-362-7163	

Neurosurgery	Dr. Michael Chicoine	660 S. Euclid Ave	314-362-4313	
Plastic surgery	Dr. Susan Mackinnon Dr. Albert Woo	660 S. Euclid Ave	314-362-4586	
Neuro-oncology	Dr. Gerald Linette	660 S. Euclid Ave	314-362-5677	
Medical Oncology/Radiation Oncology	Dr. Joseph Simpson	660 S. Euclid Ave	314-362-8567	
Audiology/ENT	Dr. Timothy Hullar	660 S. Euclid Ave	314-362-8641	
Radiology/ Neuroradiology	Dr. Robert McKinstry	#1 Children's Place	314-454-2525	
General Surgery/Surgical Oncology	Dr. Jeffrey Moley	660 S. Euclid Ave	314-747-0064	
Dermatology	Dr. Roberta Sengelmann	660 S. Euclid Ave	314-996-8824	
Cardiovascular Disease	Dr. Alan Braverman	660 S. Euclid Ave	314-362-1291	
Oral and Maxillofacial Surgery	Dr. Mark Beehner	660 S. Euclid Ave	314-362-4559	
Pain	Dr. Robert Swarm	660 S. Euclid Ave	314-362-8820	

4. NUMBER OF NF PATIENTS YOUR CLINIC SEES

- a. How many NF PATIENTS did you see in the past 12 months?
- b. How many of these were NEW patients to your clinic?

*Insert numbers below*

	NF1	NF2	SCHWANNOMATOSIS	OTHER
NUMBER OF PATIENTS SEEN IN PAST 12 MONTHS	229	0	0	0
NUMBER OF <u>NEW</u> PATIENTS SEEN IN PAST 12 MONTHS	75	0	0	0
TOTAL	229	0	0	0

- c. Overall what proportion of patients seen in the past year were (give finite numbers if these are available, or estimate percentage):

Under 18 32      18+ 197      (give numbers - if data available)

## 5. TRANSITIONING PEDIATRIC TO ADULT NF CARE

How does your clinic facilitate continuity of care for patients transiting from pediatric to adult care?

*The Washington University NF Clinic operates at St. Louis Children's Hospital. We care for patients of all ages. Both adults and children are seen at St. Louis Children's Hospital. In the event of hospitalization, they are cared for by specialists in either adult or pediatric medicine.*

Explain how continuity of care is accomplished. Describe those partnering clinics with which you coordinate services, and explain any limitations:

*See above.*

## 6. INTERNAL CONFERENCES

Provide details on internal conferences in your institution which are related to NF patient care in your clinic (e.g. NF Clinic case management conference, etc.)

*The Washington University NF Clinic participates in a joint patient care conference with Neuro-Oncology. Patients cared for jointly by adult or pediatric neuro-oncology are discussed in the context of their respective tumor boards with our full participation.*

## 7. CLINICAL TRIALS

Our clinic is willing and able to provide our NF patients with information on, and to facilitate their participation in, clinical trials for which NF patients are eligible (check box)

X Yes                       No

If 'no', briefly describe why.

Do you currently refer patients to clinical trials?

X Yes                       No

If 'yes', provide details of current clinical trial protocols in which you currently or have had patients involved in the past 5 years.

*We have enrolled patients with plexiform neurofibromas in the Perfenidone, farnesyltransferase, and AZD clinical trials.*

## 8. PATIENT REGISTRY

Do you currently have an NF specific patient database/registry?

Yes  No

If 'yes', please describe.

Would you be willing to transfer this data to a centralized CTF NF Database?

Yes  No

If 'no', explain your limitations.

## 9. PUBLICATIONS and RESEARCH (IF APPLICABLE)

a. Please list any relevant NF publications from your clinic in the past 5 years. Include Journal, Citation and Title.

1. Chan CC, Koch CA, Kaiser-Kupfer MI, Parry DM, Gutmann DH, Zhuang Z, Vortmeyer AO: Loss of heterozygosity for the NF2 gene in retinal and optic nerve lesions of patients with neurofibromatosis 2. *J. Pathol.* 198:14-20, 2002.
2. Chan CC, Koch CA, Kaiser-Kupfer MI, Parry DM, Gutmann DH, Zhuang Z, Vortmeyer AO: Loss of heterozygosity for the NF2 gene in retinal and optic nerve lesions of patients with neurofibromatosis 2. *J. Pathol.* 198:14-20, 2002.
3. Friedman JM, Arbiser J, Epstein JA, Gutmann DH, Huot SJ, Lin A, McManus B, Korf BR: Cardiovascular disease in neurofibromatosis 1: Report of the NF1 cardiovascular task force. *Genetics in Medicine* 4:105-11, 2002.
4. Packer RJ, Gutmann DH, Rubenstein A, Viskochil D, Zimmerman RA, Vezina G, Small J, Korf B: Plexiform neurofibromas in NF1: Towards biologic-based therapy. *Neurology* 58:1461-1470, 2002.
5. Baser ME, De Rienzo A, Altomare D, Balsara BR, Hedrick NM, Gutmann DH, Pitts LH, Jackler RK, Testa JR. Neurofibromatosis 2 and malignant mesothelioma. *Neurology* 59:290-291, 2002.
6. Perry A, Kunz SN, Fuller CE, Banerjee R, Marley EF, Liapis H, Watson MA, Gutmann DH. Differential NF1, p16, and EGFR patterns by interphase cytogenetics (FISH) in malignant peripheral nerve sheath tumor (MPNST) and morphologically similar spindle cell neoplasms. *J. Neuropathol Exp Neurol.* 61:702-9, 2002.
7. Gutmann DH, Rasmussen SA, Wolkenstein P, MacCollin MM, Guha A, Inskip PD, North KN, Poyhonen M, Birch PH, Friedman JM. Gliomas presenting after age ten in individuals with neurofibromatosis type 1 (NF1). *Neurology* 59:759-761, 2002.
8. King A, Listernick R, Charrow J, Piersall L, Gutmann DH: Optic pathway gliomas in neurofibromatosis type 1: The effect of presenting symptoms on outcome. *American Journal of Medical Genetics* 122:95-99, 2003.
9. Gutmann DH, James CD, Poyhonen M, Louis DN, Ferner R, Guha A, Hariharan S, Viskochil DV, Perry A: Molecular analysis of astrocytomas presenting after age 10 in individuals with NF1. *Neurology* 61:1397-1400, 2003.
10. Spritz RA, Itin PH, Gutmann DH: Piebaldism and Neurofibromatosis Type 1: Horses of Very Different Colors. *J. Invest. Dermatol.* 122:xxxiv-xxxv, 2004.

11. Fedi M, Mitchell LA, Renate M Kalnins RM, Gutmann DH, Perry A, Newton M, Brodtmann A, Berkovic SF: *Glioneuronal Tumors in Neurofibromatosis Type 1: MRI-Pathological Study*. *J Clin. Neurosci.* 11:745-7, 2004.
12. Watson MA, Perry A, Tihan T, Prayson RA, Guha A, Bridge J, Ferner R, Gutmann DH: *Gene expression profiling reveals unique molecular subtypes of neurofibromatosis type 1-associated and sporadic malignant peripheral nerve sheath tumors*. *Brain Pathol.* 14: 297-303, 2004.
13. Listerick R, Ferner RE, Piersall LS, Sharif S, DH Gutmann DH, Charrow J: *Late-onset optic pathway tumors in children with neurofibromatosis 1 (NF1)*. *Neurology* 63:1944-1946, 2004.
14. Miller SJ, Rangwala F, Williams J, Ackerman P, Kong S, Jegga A, Aronow B, Frahm S, Kluwe L, Mautner V, Upadhyaya M, Muir D, Wallace M, Hagen J, Quelle DE, Watson MA, Perry A, Gutmann DH, Ratner N: *Large-scale molecular comparison of human Schwann cells to malignant peripheral nerve sheath tumor cells and tissues*. *Cancer Res.* 66:2584-91, 2006.
15. Leonard JR, Perry A, Rubin JB, King AA, Chicoine MR, Gutmann DH: *The role of surgical biopsy in the diagnosis of glioma in individuals with neurofibromatosis-1*. *Neurology* 67:1509-1512, 2006.
16. Spinner RJ, Scheithauer BW, Perry A, Amrami KK, Emmett R, Gutmann DH: *Co-localized cellular schwannoma and plexiform neurofibroma in the absence of neurofibromatosis*. *J. Neurosurgery* (in press).
17. Listerick R, Ferner RE, Liu GT, Gutmann DH: *Optic pathway gliomas in neurofibromatosis-1: Controversies and recommendations*. *Annals of Neurology* 61:189-98, 2007.
18. Leonard JR, Ferner RE, Thomas N, Gutmann DH: *Cervical cord compression from plexiform neurofibromas in neurofibromatosis-1*. *Journal of Neurology, Neurosurgery, and Psychiatry* (in press).
19. Ess KC, Gutmann DH: *Neurofibromatoses*. In: *Manual of Neurological Practice*. (R. Evans, editor). WB Saunders: Orlando. Pages 896-898, 2003.
20. Lynch TM, Gutmann DH: *Neurofibromatosis 1*. In: *Neurologic Clinics of North America*. 20:841-865, 2002.
21. McCullough LA, Gutmann DH: *GTPase activating proteins: Neurofibromatosis 1*. In: *Molecular Basis of Inborn Errors of Development*. Oxford University Press: San Francisco 2003.
22. Gurnett CA, Gutmann DH: *Neuro-oncology: The Neurofibromatoses*. In: *The Molecular and Genetic Basis of Neurological and Psychiatric Disease*. 3<sup>rd</sup> Edition (Rosenberg RN, et al., editors). Butterworth & Heinemann: MA. Pages 389-398, 2003.
23. Esper GJ, Gutmann DH: *Neurofibromatosis 1 and 2*. In: *Principles of Molecular Medicine*. 2<sup>nd</sup> edition. (S. Strittmatter, et al., editors) Humana Press: NJ, 2004.
24. Gutmann DH, Wetmore C, O'Neill BP: *The Phakomatoses*. In: *Principles of Neuro-Oncology* (D. Schiff, editor) McGraw Hill: Philadelphia pp 233-256, 2005.
25. Malhotra A, Gutmann DH: *Neurofibromatosis 1*. In: *Neurogenetics: Scientific and Clinical Advances*. (D. Lynch, editor). Marcel Dekker, Inc.: New York. 2005.
26. Gutmann DH, Perry A: *Neurofibromatosis 1*. In: *Russell & Rubinstein Pathology of Tumors of the Nervous System*, 7<sup>th</sup> Edition, Hodder Arnold Press: London, Pages 903-916, 2006.
27. Leeman B, Gutmann DH: *Neurofibromatosis 1*. In: *Current Pediatric Therapy*. 18<sup>th</sup> Edition. (Burg, Ingelfinger, Polin, and Gershon, editors). Elsevier Press: Philadelphia, Pages 396-399, 2006.
28. Piersall L, Gutmann DH: *Neurofibromatosis 1*. In: *Neurobiology of Disease* (S. Gilman, editor) Elsevier: New York Pages 413-424, 2006.
29. Daginakatte GC, Gutmann DH: *NF1 and Neurofibromatosis 1*. In: *Molecular Basis of Inborn Errors of Development*. Oxford University Press: San Francisco. 2<sup>nd</sup> edition (in press).

30. Bogitch R, Gutmann DH: *Neuro-oncology: The Neurofibromatoses*. In: *The Molecular and Genetic Basis of Neurological and Psychiatric Disease*. 4<sup>th</sup> Edition (Rosenbery RN, et al., editors). Butterworth & Heinemann: MA. (in press).
31. Jost S, Gutmann DH: *Neurofibromatosis and Other Genetic Syndromes*. In: *Handbook of Clinical Neurology, Neuro-Oncology Volume*. (Aminoff, Biller and Swaab, editors). Elsevier: New York (in press).
32. Piersall L, Gutmann DH, Ferner R: *Living with neurofibromatosis type 1: A guide for adults*. National Neurofibromatosis Foundation, 2002.
33. Piersall L, Gutmann DH: *Let's Talk About Neurofibromatosis: A guide for teens*. Washington University, 2006.
34. Piersall L, Gutmann DH: *Understanding Your Child's Diagnosis of Neurofibromatosis*. Washington University, 2007.

b. Please provide information on NF-related research ongoing in your clinic or performed by personnel affiliated with your clinic.

*The Washington University NF Clinic is embedded within the Washington University NF Center of Excellence. The NF Center was established to position Washington University as an international leader in NF basic and clinical research as well as to serve as a beacon for "bench to bedside" research in NF and related nervous system tumors. Using a team approach to gain a complete understanding of the complexities of NF, the mission of the Washington University NF Center is focused on:*

- *Promoting pioneering laboratory research aimed at understanding the roles of the NF genes in health and disease*
- *Facilitating collaborative interdisciplinary basic science and clinical research in NF*
- *Translating innovative scientific discoveries to improved care for individuals with NF*

*More information can be found on our website at <http://neuro.wustl.edu/nfcenter>.*

## 10. PATIENT SUPPORT

Do you have an NF patient support group that meets in association with your NF Clinic?

*Not currently, but we are interested in strengthening our relationship with the local community. We are currently working with the Children's Tumor Foundation to establish these group meetings.*