

Neurofibromatosis Research Program

Research Resources Index



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Neurofibromatosis Research Program (NFRP) Research Resources Index

The NFRP now offers a service on our website featuring a list of available NF research resources with a goal to facilitate and speed NF research by publicizing new resources and aiding collaborations. In addition to providing access to resources developed through our program, Principal Investigators (PIs) can contact other sites, repositories, or the PI directly for information and requests.

36% of total funds awarded
between FY99-FY16
(\$95,027,848)
have resulted in
resource development



118
Total Resources
Developed Resulting from
102
Funded Awards



**NFRP
Research
Resources**

**Drosophila
Models**

**Mouse
Models**

**Zebrafish
Models**

Antibodies

**Yeast
Strains**

**Rat
Models**

**Cell and
Molecular
Bio Tools**

Methods


**Databases
& Data
Sets**

**Cell
Lines**

Animal Models

An animal model is a living, non-human organism used for research for the purpose of understanding the human disease process without the added risk of human involvement and harm. Commonly used animals for this purpose include mice and rats. In addition to these, the NFRP research resources index includes drosophila and zebrafish models. The study of the NF disease process including possible causes, the conditions that can promote or hinder pathogenesis and other factors in different animal models that have similarities with the human body is very useful to better understand how the disease evolves in humans and can lead to identifying different avenues for treatment, control and prevention.

DROSOPHILA MODELS

Award Number	Principal Investigator Award Amount	Products or Resources	References
 <p>NF000002</p> <p>Click on Any Award Number for More Information</p>	<p>Dennis LaJeunesse \$371,050.00</p>	<ul style="list-style-type: none"> • Creation of eight FLAG versions of scribbler (two wild-type controls) versions and six deletion mutants named: FLAGSbbA, FLAGSbbB, FLAGSbbA::A., FLAGSbbB::A., FLAGSbbB::Zf, FLAGSbbB::B, FLAGSbbAZfB C-term truncation, and FLAGSbbZfB C-term truncation. • Five new strains of Drosophila carrying mutations in the scribbler gene. These strains will be kept in the laboratory and will be available to all researchers once they have been fully characterized and published. 	<ol style="list-style-type: none"> 1. LaJeunesse DR, Buckner SM, Lake J, Na C, Pirt A, Fromson K. 2004. Three new Drosophila markers of intracellular membranes. <i>Biotechniques</i>. May;36(5):784-8, 790. doi: 10.2144/04365ST01. PMID: 15152597. 2. LaJeunesse D. 2010. Alteration of wing size through over-expression of scribbler isoforms. <i>Fly (Austin)</i>. Oct-Dec;4(4):320-6. doi: 10.4161/fly.4.4.13148. Epub 2010 Oct 1. PMID: 20798603.
<p>NF030006</p>	<p>Michael Stern \$1,209,480.79</p>	<ul style="list-style-type: none"> • D42>PI3K-CAAX • DmGluRA112b;D42>PI3K-CAAX • D42>PI3KDN • Foxo21/Foxo25 • Foxo21/Foxo25;OK6>PI3KDN • D42>PI3K-CAAX;Foxo+ • OK6>PI3KDN • D42>Foxo+ • D42>PI3K-CAAX;S6KDN • D42>S6KACT • D42>S6KDN • gli>RasV12 • PI3K2H1;gli>RasV12 • gli>PI3KDN,RasV12 • gli>PI3K-CAAX • PI3K2H1/PI3KA;gli>PI3K-CAAX • MZ709>PI3K-CAAX • gli-RafF179 • gli>RalV20 • Akt4226;gli>PI3K-CAAX • Akt4226/+;gli>PI3K-CAAX • gli>PI3K-CAAX;Foxo+(f19-5) • gli>PI3K-CAAX, Foxo+(m3-1) • gli>PI3K-CAAX, GFP 	<ol style="list-style-type: none"> 1. Howlett E, Lin CC, Lavery W, Stern M. 2008. A PI3-kinase-mediated negative feedback regulates neuronal excitability. <i>PLoS Genet</i>. Nov;4(11):e1000277. doi: 10.1371/journal.pgen.1000277. Epub 2008 Nov 28. PMID: 19043547; PMCID: PMC2581892. 2. Lavery W, Hall V, Yager JC, Rottgers A, Wells MC, Stern M. 2007. Phosphatidylinositol 3-kinase and Akt nonautonomously promote perineurial glial growth in Drosophila peripheral nerves. <i>J Neurosci</i>. Jan 10;27(2):279-88. doi: 10.1523/JNEUROSCI.3370-06.2007. PMID: 17215387; PMCID: PMC6672080.

DROSOPHILA MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF080012	Michael Stern \$138,211.00	<ul style="list-style-type: none"> D42>CaMKIIT287D D42>CaMKIIT287A D42>ala DFaKCG1,D42>DFaK+ DFaKCG1, D42>CaMKIIT287D D42>PI3KDN, CaMKIIT287D D42>PI3K-CAAX, ala DFaKCG1,D42>PI3K-CAAX DmGluRA112b;D42>CaMKIIT287D DmGluRA112b; D42>+ 	<ol style="list-style-type: none"> Chun-Jen Lin C, Summerville JB, Howlett E, Stern M. 2011. The metabotropic glutamate receptor activates the lipid kinase PI3K in Drosophila motor neurons through the calcium/calmodulin-dependent protein kinase II and the nonreceptor tyrosine protein kinase DFak. <i>Genetics</i>. Jul;188(3):601-13. doi: 10.1534/genetics.111.128561. Epub 2011 Apr 21. PMID: 21515581; PMCID: PMC3176550.
NF080075 (NF073069)	James Walker \$662,251.00	<p>Developed a Drosophila model for schwannomatosis Snr101319 homozygous Drosophila expressing:</p> <ul style="list-style-type: none"> actin5C-GAL4>SMARCB1-a actin5C-GAL4>SMARCB1-B actin5C-GAL4>SMARCB1 P14H actin5C-Gal4>SMARCB1 R53L actin5C-GAL4>SMARCB1 ΔExon 4 	<ol style="list-style-type: none"> Di Stefano L, Walker JA, Burgio G, Corona DF, Mulligan P, Näär AM, Dyson NJ. 2011. Functional antagonism between histone H3K4 demethylases in vivo. <i>Genes Dev</i>. Jan 1;25(1):17-28. doi: 10.1101/gad.1983711. PMID: 21205864; PMCID: PMC3012933. Miles WO, Dyson NJ, Walker JA. 2011. Modeling tumor invasion and metastasis in Drosophila. <i>Dis Model Mech</i>. Nov;4(6):753-61. doi: 10.1242/dmm.006908. Epub 2011 Oct 6. PMID: 21979943; PMCID: PMC3209645. Longworth MS, Walker JA, Anderssen E, Moon NS, Gladden A, Heck MM, Ramaswamy S, Dyson NJ. 2012. A shared role for RBF1 and dCAP-D3 in the regulation of transcription with consequences for innate immunity. <i>PLoS Genet</i>. 8(4):e1002618. doi: 10.1371/journal.pgen.1002618. Epub 2012 Apr 4. PMID: 22496667; PMCID: PMC3320600. Smith MJ, Walker JA, Shen Y, Stemmer-Rachamimov A, Gusella JF, Plotkin SR. 2012. Expression of SMARCB1 (INI1) mutations in familial schwannomatosis. <i>Hum Mol Genet</i>. Dec 15;21(24):5239-45. doi: 10.1093/hmg/dds370. Epub 2012 Sep 4. PMID: 22949514; PMCID: PMC3510754.

MOUSE MODELS

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF000004	Faith Strickland \$463,937.34	<ul style="list-style-type: none"> Nf1+;/+;[Tg]Dct:lacZC57BL/6J mice 	
NF010016	Marco Giovannini and David Gutmann \$785,230.00	<ul style="list-style-type: none"> Development of Protein 4.1B knockout mice. Development of Protein 4.1B conditional knockout mice. Development of tumor suppressor in lung cancer-1 knockout mice. 	<ol style="list-style-type: none"> Surace EI, Lusic E, Murakami Y, Scheithauer BW, Perry A, Gutmann DH. 2004. Loss of tumor suppressor in lung cancer-1 (TSLC1) expression in meningioma correlates with increased malignancy grade and reduced patient survival. <i>J Neuropathol Exp Neuro</i>. Oct;63(10):1015-27. doi: 10.1093/jnen/63.10.1015. PMID: 15535129. McClatchey AI, Giovannini M. 2005. Membrane organization and tumorigenesis--the NF2 tumor suppressor, Merlin. <i>Genes Dev</i>. Oct 1;19(19):2265-77. doi: 10.1101/gad.1335605. PMID: 16204178."

Animal Models

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF010054	Nancy Ratner \$706,506.00	<ul style="list-style-type: none"> Development of a transgenic mouse strain expressing wild-type human epidermal growth factor receptor under the control of the Schwann cell 2',3'-cyclic nucleotide 3'-phosphodiesterase promoter. CNP-hEGFR transgenic mouse model. NF1+/-; p53+/-; EGFRwa-2/+ mouse model system. Transgenic mice expressing human epidermal growth factor receptor under the control of the Schwann cell 2',3'-cyclic nucleotide 3'-phosphodiesterase promoter (CNP-hEGFR mice). 	<ol style="list-style-type: none"> Aguirre A, Rizvi TA, Ratner N, Gallo V. Overexpression of the epidermal growth factor receptor confers migratory properties to nonmigratory postnatal neural progenitors. <i>J Neurosci.</i> Nov 30;25(48):11092-106. doi: 10.1523/JNEUROSCI.2981-05.2005. PMID: 16319309; PMCID: PMC6725641. Wu J, Crimmins JT, Monk KR, Williams JP, Fitzgerald ME, Tedesco S, Ratner N. 2006. Perinatal epidermal growth factor receptor blockade prevents peripheral nerve disruption in a mouse model reminiscent of benign world health organization grade I neurofibroma. <i>Am J Pathol.</i> May;168(5):1686-96. doi: 10.2353/ajpath.2006.050859. PMID: 16651634; PMCID: PMC1606591.
NF010093 (NF990018)	Kevin Shannon \$4,169,173.00	<ul style="list-style-type: none"> Model of myeloproliferative disorder: Mx1-Cre; Nf1 flox/flox 	<ol style="list-style-type: none"> Crone SA, Negro A, Trumpp A, Giovannini M, Lee KF. 2003. Colonic epithelial expression of ErbB2 is required for postnatal maintenance of the enteric nervous system. <i>Neuron.</i> Jan 9;37(1):29-40. doi: 10.1016/s0896-6273(02)01128-5. PMID: 12526770. Denisenko-Nehrbass N, Goutebroze L, Galvez T, Bonnon C, Stankoff B, Ezan P, Giovannini M, Faivre-Sarrailh C, Girault JA. 2003. Association of Caspr/paranodin with tumour suppressor schwannomin/merlin and beta1 integrin in the central nervous system. <i>J Neurochem.</i> Jan;84(2):209-21. doi: 10.1046/j.1471-4159.2003.01503.x. PMID: 12558984. Fleury-Feith J, Lecomte C, Renier A, Matrat M, Kheuang L, Abramowski V, Levy F, Janin A, Giovannini M, Jaurand MC. 2003. Hemizyosity of Nf2 is associated with increased susceptibility to asbestos-induced peritoneal tumours. <i>Oncogene.</i> Jun 12;22(24):3799-805. doi: 10.1038/sj.onc.1206593. PMID: 12802287.*

*Additional references can be found in Appendix A.

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF020005	David Gutmann \$1,448,199.00	<ul style="list-style-type: none"> Developed first published model of NF1-associated optic nerve glioma. Dasgupta B. 2005. Development of the second published mouse strain. Full details of this mouse strain are provided in Appendix, Manuscript #1. Mouse model of NF1-associated optic nerve glioma. The full details of this mouse strain are provided in Bajenaru ML, Hernandez MR, Perry A, et al. 2003. Optic nerve glioma in mice requires astrocyte Nf1 gene inactivation and Nf1 brain heterozygosity. <i>Cancer Res</i> 63:8573-8577. In collaboration with Dr. Luis Parada, developed another published model of NF1-associated optic nerve glioma. The full details of this mouse strain are provided in Zhu Y, Harada T, Liu L, et al. 2005. Inactivation of NF1 in CNS causes increased glial progenitor proliferation and optic glioma formation. <i>Development</i> 132:5577-5588. Developed transgenic mice that express Cre recombinase in progenitor cells (neural stem cells) beginning at embryonic day 9 (BLBP-Cre mice). Developed LSL-Rheb mice that can conditionally express Rheb upon Cre expression. 	<ol style="list-style-type: none"> Dasgupta B, Dugan LL, Gutmann DH. 2003. The neurofibromatosis 1 gene product neurofibromin regulates pituitary adenylate cyclase-activating polypeptide-mediated signaling in astrocytes. <i>J Neurosci</i>. Oct 1;23(26):8949-54. doi: 10.1523/JNEUROSCI.23-26-08949.2003. PMID: 14523097; PMCID: PMC6740397. Bajenaru ML, Hernandez MR, Perry A, Zhu Y, Parada LF, Garbow JR, Gutmann DH. 2003. Optic nerve glioma in mice requires astrocyte Nf1 gene inactivation and Nf1 brain heterozygosity. <i>Cancer Res</i>. Dec 15;63(24):8573-7. PMID: 14695164. Dasgupta B, Li W, Perry A, Gutmann DH. 2005. Glioma formation in neurofibromatosis 1 reflects preferential activation of K-RAS in astrocytes. <i>Cancer Res</i>. Jan 1;65(1):236-45. PMID: 15665300.*
NF020061	Stefan Mundlos \$405,172.00	<ul style="list-style-type: none"> Nf1flox/Nf1flox/Prx1Cre mice. 	
NF030020	David Gutmann \$1,183,129.00	<ul style="list-style-type: none"> Developed several founder lines of Tslc1+/- mice. 	<ol style="list-style-type: none"> Surace EI, Strickland A, Hess RA, Gutmann DH, Naughton CK. 2006. Tslc1 (nectin-like molecule-2) is essential for spermatozoa motility and male fertility. <i>J Androl</i>. Nov-Dec;27(6):816-25. doi: 10.2164/jandrol.106.000398. Epub 2006 Jul 12. PMID: 16837733; PMCID: PMC2755522. Houshmandi SS, Surace EI, Zhang HB, Fuller GN, Gutmann DH. 2006. Tumor suppressor in lung cancer-1 (TSLC1) functions as a glioma tumor suppressor. <i>Neurology</i>. Nov 28;67(10):1863-6. doi: 10.1212/01.wnl.0000244472.56198.84. PMID: 17130425. Chadee DN, Xu D, Hung G, Andalibi A, Lim DJ, Luo Z, Gutmann DH, Kyriakis JM. 2006. Mixed-lineage kinase 3 regulates B-Raf through maintenance of the B-Raf/Raf-1 complex and inhibition by the NF2 tumor suppressor protein. <i>Proc Natl Acad Sci U S A</i>. Mar 21;103(12):4463-8. doi: 10.1073/pnas.0510651103. Epub Mar 13. PMID: 16537381; PMCID: PMC1450194.*

*Additional references can be found in Appendix A.

Animal Models

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF043015	Stefan Pulst \$1,035,244.00	<ul style="list-style-type: none"> Hrs +/+; Nf2 +/+ , Hrs +/+; Nf2 +/- , Hrs +/-; Nf2 +/+ , Hrs +/-; Nf2 +/- mice have been produced. New mouse lines have been generated that contain Hrs and Nf2 null alleles either in trans or in cis. 	
NF043019	David Ingram \$677,250.00	<ul style="list-style-type: none"> Nf1 +/-; Tie2-GFP mice p85&#945; +/-; Tie2-GFP mice 	<ol style="list-style-type: none"> Munchhof AM, Li F, White HA, Mead LE, Krier TR, Fenoglio A, Li X, Yuan J, Yang FC, Ingram DA. 2006. Neurofibroma-associated growth factors activate a distinct signaling network to alter the function of neurofibromin-deficient endothelial cells. <i>Hum Mol Genet.</i> Jun 1;15(11):1858-69. doi: 10.1093/hmg/ddl108. Epub 2006 Apr 28. PMID: 16648142. Wu X, Estwick SA, Chen S, Yu M, Ming W, Nebesio TD, Li Y, Yuan J, Kapur R, Ingram D, Yoder MC, Yang FC. 2006. Neurofibromin plays a critical role in modulating osteoblast differentiation of mesenchymal stem/progenitor cells. <i>Hum Mol Genet.</i> Oct 1;15(19):2837-45. doi: 10.1093/hmg/ddl208. Epub 2006 Aug 7. PMID: 16893911. Yang FC, Chen S, Robling AG, Yu X, Nebesio TD, Yan J, Morgan T, Li X, Yuan J, Hock J, Ingram DA, Clapp DW. 2006. Hyperactivation of p21ras and PI3K cooperate to alter murine and human neurofibromatosis type 1-haploinsufficient osteoclast functions. <i>J Clin Invest.</i> Nov;116(11):2880-91. doi: 10.1172/JCI29092. Epub 2006 Oct 19. PMID: 17053831; PMCID: PMC1616197.*
NF043040	Andrea McClatchey \$830,034.00	<ul style="list-style-type: none"> Model of renal carcinoma: Vil-Cre; Nf2 lox/lox 	<ol style="list-style-type: none"> Curto M, Cole BK, Lallemand D, Liu CH, McClatchey AI. 2007. Contact-dependent inhibition of EGFR signaling by Nf2/Merlin. <i>J Cell Biol.</i> Jun 4;177(5):893-903. doi: 10.1083/jcb.200703010. PMID: 17548515; PMCID: PMC2064288. Cole BK, Curto M, Chan AW, McClatchey AI. 2008. Localization to the cortical cytoskeleton is necessary for Nf2/merlin-dependent epidermal growth factor receptor silencing. <i>Mol Cell Biol.</i> Feb;28(4):1274-84. doi: 10.1128/MCB.01139-07. Epub 2007 Dec 17. PMID: 18086884; PMCID: PMC2258752. Curto M, McClatchey AI. 2008. Nf2/Merlin: a coordinator of receptor signalling and intercellular contact. <i>Br J Cancer.</i> Jan 29;98(2):256-62. doi: 10.1038/sj.bjc.6604002. Epub 2007 Oct 30. PMID: 17971776; PMCID: PMC2361439.*
NF043050 (NF010093)	Kevin Shannon \$4,501,783.00	<ul style="list-style-type: none"> Paternal inheritance of Nestin-Cre resulting in mosaic loss of Nf2: NesCreP; Nf2 flox/flox 	<ol style="list-style-type: none"> Bonilha VL, Rayborn ME, Saotome I, McClatchey AI, Hollyfield JG. 2006. Microvilli defects in retinas of ezrin knockout mice. <i>Exp Eye Res.</i> Apr;82(4):720-9. doi: 10.1016/j.exer.2005.09.013. Epub 2005 Nov 10. PMID: 16289046. Parada LF, Kwon CH, Zhu Y. 2005. Modeling neurofibromatosis type 1 tumors in the mouse for therapeutic intervention. <i>Cold Spring Harb Symp Quant Biol.</i> 70:173-6. doi: 10.1101/sqb.2005.70.025. PMID: 16869751. Yang FC, Chen S, Clegg T, Li X, Morgan T, Estwick SA, Yuan J, Khalaf W, Burgin S, Travers J, Parada LF, Ingram DA, Clapp DW. 2006. Nf1 +/- mast cells induce neurofibroma like phenotypes through secreted TGF-beta signaling. <i>Hum Mol Genet.</i> Aug 15;15(16):2421-37. doi: 10.1093/hmg/ddl165. Epub 2006 Jul 11. PMID: 16835260; PMCID: PMC3024714.*

*Additional references can be found in Appendix A.

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF043126</u>	Evelyn Tiffany-Castiglioni \$99,867.19	<ul style="list-style-type: none"> Breeding colony of mice to produce offspring heterozygous for an NF1 mutation. 	
<u>NF050035</u>	Nancy Ratner \$1,070,855.00	<ul style="list-style-type: none"> Nuclear factor 1a (NF1a) Plp-Cre+;Nfl flox/flox knockout Nfl flox/flox male mice Plp-Cre+ female mice lox-stop-lox b-actin-EGFP+ plp-Cre+ Nf1 fl/fl mouse model Development of Plp-Cre;Nfl fl/fl model. 	
<u>NF050041</u>	Yuan Zhu \$1,462,840.00	<ul style="list-style-type: none"> p53CK01 (p53+/-;NF1-/flox;POA-cre+;R26R-LacZ/+) p53CK02 (p53+/-;NF1flox/flox;POAcre+;R26R-LacZ/+) 	<ol style="list-style-type: none"> Joseph NM, Mosher JT, Buchstaller J, Snider P, McKeever PE, Lim M, Conway SJ, Parada LF, Zhu Y, Morrison SJ. 2008. The loss of Nf1 transiently promotes self-renewal but not tumorigenesis by neural crest stem cells. <i>Cancer Cell</i>. Feb;13(2):129-40. doi: 10.1016/j.ccr.2008.01.003. PMID: 18242513; PMCID: PMC2566828. Roth TM, Ramamurthy P, Muir D, Wallace MR, Zhu Y, Chang L, Barald KF. 2008. Influence of hormones and hormone metabolites on the growth of Schwann cells derived from embryonic stem cells and on tumor cell lines expressing variable levels of neurofibromin. <i>Dev Dyn</i>. Feb;237(2):513-24. doi: 10.1002/dvdy.21430. PMID: 18213578. Zheng H, Chang L, Patel N, Yang J, Lowe L, Burns DK, Zhu Y. 2008. Induction of abnormal proliferation by nonmyelinating schwann cells triggers neurofibroma formation. <i>Cancer Cell</i>. Feb;13(2):117-28. doi: 10.1016/j.ccr.2008.01.002. PMID: 18242512.
<u>NF050145</u>	Victor-Felix Mautner \$628,550.00	<ul style="list-style-type: none"> MPNST S462 and S805 mouse xenograft model PNF xenograft model 	<ol style="list-style-type: none"> Demestre M, Messerli SM, Celli N, Shahhossini M, Kluwe L, Mautner V, Maruta H. 2009. CAPE (caffeic acid phenethyl ester)-based propolis extract (Bio 30) suppresses the growth of human neurofibromatosis (NF) tumor xenografts in mice. <i>Phytother Res</i>. Feb;23(2):226-30. doi: 10.1002/ptr.2594. PMID: 18726924.
<u>NF060106</u>	Miguel Sena-Esteves \$110,192.00	<ul style="list-style-type: none"> NF2flox2/flox2FLucflox mice 	

Animal Models

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF073112	Feng-Chun Yang \$772,870.00	<ul style="list-style-type: none"> NF1 skeletal model with reduced bone mineral density, reduced calcium ossification, reduced bone turnover, and increased ratio of spinal canal area to vertebral body area 	<ol style="list-style-type: none"> Wu X, Chen S, He Y, Rhodes SD, Mohammad KS, Li X, Yang X, Jiang L, Nalepa G, Snider P, et al. 2011. The haploinsufficient hematopoietic microenvironment is critical to the pathological fracture repair in murine models of neurofibromatosis type 1. <i>PLoS One</i>. 6(9):e24917. doi: 10.1371/journal.pone.0024917. Epub 2011 Sep 29. PMID: 21980365; PMCID: PMC3182976. Sharma R, Wu X, Rhodes SD, Chen S, He Y, Yuan J, Li J, Yang X, Li X, Jiang L, et al. 2013. Hyperactive Ras/MAPK signaling is critical for tibial nonunion fracture in neurofibromin-deficient mice. <i>Hum Mol Genet</i>. Dec 1;22(23):4818-28. doi: 10.1093/hmg/ddt333. Epub 2013 Jul 17. PMID: 23863460; PMCID: PMC3820137. Ferguson MJ, Rhodes SD, Jiang L, Li X, Yuan J, Yang X, Zhang S, Vakili ST, Territo P, Hutchins G, et al. 2016. Preclinical Evidence for the Use of Sunitinib Malate in the Treatment of Plexiform Neurofibromas. <i>Pediatr Blood Cancer</i>. Feb;63(2):206-13. doi: 10.1002/pbc.25763. Epub 2015 Sep 16. PMID: 26375012; PMCID: PMC4862309.
NF080045	Kevin Pumiglia \$824,085.12	<ul style="list-style-type: none"> Inducible knockdown of Nf1 in the vascular endothelium: CAD5-CreERT2/Rosa26-LSL-td-Tomato/NF1flox/flox 	<ol style="list-style-type: none"> Bajaj A, Li QF, Zheng Q, Pumiglia K. 2012. Loss of NF1 expression in human endothelial cells promotes autonomous proliferation and altered vascular morphogenesis. <i>PLoS One</i>. 7(11):e49222. doi: 10.1371/journal.pone.0049222. Epub 2012 Nov 7. PMID: 23145129; PMCID: PMC3492274.
NF093053	Nancy Ratner \$984,663.00	<ul style="list-style-type: none"> PlpCre;Nf1 flox/flox 	<ol style="list-style-type: none"> Kalamarides M, Acosta MT, Babovic-Vuksanovic D, Carpen O, Cichowski K, Evans DG, Giancotti F, Hanemann CO, Ingram D, Lloyd AC, Mayes DA, Messiaen L, Morrison H, North K, Packer R, Pan D, Stemmer-Rachamimov A, Upadhyaya M, Viskochil D, Wallace MR, Hunter-Schaedle K, Ratner N. 2012. Neurofibromatosis 2011: a report of the Children's Tumor Foundation annual meeting. <i>Acta Neuropathol</i>. Mar;123(3):369-80. doi: 10.1007/s00401-011-0905-0. Epub 2011 Nov 16. PMID: 22083253; PMCID: PMC3282898. Mayes DA, Rizvi TA, Cancelas JA, Kolasinski NT, Ciraolo GM, Stemmer-Rachamimov AO, Ratner N. 2011. Perinatal or adult Nf1 inactivation using tamoxifen-inducible PlpCre each cause neurofibroma formation. <i>Cancer Res</i>. Jul 1;71(13):4675-85. doi: 10.1158/0008-5472.CAN-10-4558. Epub 2011 May 6. PMID: 21551249; PMCID: PMC3464476. Mayes DA, Rizvi TA, Titus-Mitchell H, Oberst R, Ciraolo GM, Vorhees CV, Robinson AP, Miller SD, Cancelas JA, Stemmer-Rachamimov AO, Ratner N. 2013. Nf1 loss and Ras hyperactivation in oligodendrocytes induce NOS-driven defects in myelin and vasculature. <i>Cell Rep</i>. Sep 26;4(6):1197-212. doi: 10.1016/j.celrep.2013.08.011. Epub 2013 Sep 12. PMID: 24035394; PMCID: PMC3982616.*
NF093074	Slawomir Antoszczyk \$173,697.59	<ul style="list-style-type: none"> MPNST sciatic nerve sheath tumor model in immune competent mice 	<ol style="list-style-type: none"> Antoszczyk S, Spyra M, Mautner VF, Kurtz A, Stemmer-Rachamimov AO, Martuza RL, Rabkin SD. Treatment of orthotopic malignant peripheral nerve sheath tumors with oncolytic herpes simplex virus. <i>Neuro Oncol</i>. 2014 Aug;16(8):1057-66. doi: 10.1093/neuonc/not317. Epub 2014 Jan 26. PMID: 24470552; PMCID: PMC4096170.

*Additional references can be found in Appendix A.

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF100136	Yuan Zhu \$1,049,181.72	<ul style="list-style-type: none"> Series of mouse models with a combination of Fbxw7 and Nf1 mutations 	<ol style="list-style-type: none"> Tan M, Zhao Y, Kim SJ, Liu M, Jia L, Saunders TL, Zhu Y, Sun Y. 2011. SAG/RBX2/ROC2 E3 ubiquitin ligase is essential for vascular and neural development by targeting NF1 for degradation. <i>Dev Cell</i>. Dec 13;21(6):1062-76. doi: 10.1016/j.devcel.2011.09.014. Epub 2011 Nov 23. PMID: 22118770; PMCID: PMC3241850. Wang Y, Kim E, Wang X, Novitch BG, Yoshikawa K, Chang LS, Zhu Y. 2012. ERK inhibition rescues defects in fate specification of Nf1-deficient neural progenitors and brain abnormalities. <i>Cell</i>. Aug 17;150(4):816-30. doi: 10.1016/j.cell.2012.06.034. PMID: 22901811; PMCID: PMC3427010. Kim E, Wang Y, Kim SJ, Bornhorst M, Jecrois ES, Anthony TE, Wang C, Li YE, Guan JL, Murphy GG, Zhu Y. 2014. Transient inhibition of the ERK pathway prevents cerebellar developmental defects and improves long-term motor functions in murine models of neurofibromatosis type 1. <i>Elife</i>. Dec 23;3:e05151. doi: 10.7554/eLife.05151. PMID: 25535838; PMCID: PMC4297949.
NF110047	Lu Le \$632,348.15	<ul style="list-style-type: none"> Neurofibroma model in a 3-D in vitro skin model mouse 	<ol style="list-style-type: none"> Chen Z, Pradhan S, Liu C, Le LQ. 2012. Skin-derived precursors as a source of progenitors for cutaneous nerve regeneration. <i>Stem Cells</i>. Oct;30(10):2261-70. doi: 10.1002/stem.1186. PMID: 22851518; PMCID: PMC3448815. Mo W, Chen J, Patel A, Zhang L, Chau V, Li Y, Cho W, Lim K, Xu J, Lazar AJ, et al. 2013. CXCR4/CXCL12 mediate autocrine cell-cycle progression in NF1-associated malignant peripheral nerve sheath tumors. <i>Cell</i>. Feb 28;152(5):1077-90. doi: 10.1016/j.cell.2013.01.053. Epub 2013 Feb 21. PMID: 23434321; PMCID: PMC3594500. Chen Z, Liu C, Patel AJ, Liao CP, Wang Y, Le LQ. 2014. Cells of origin in the embryonic nerve roots for NF1-associated plexiform neurofibroma. <i>Cancer Cell</i>. Nov 10;26(5):695-706. doi: 10.1016/j.ccell.2014.09.009. Epub 2014 Oct 30. PMID: 25446898; PMCID: PMC4254535.*
NF130064	Aaron Schindeler \$87,253.00	<ul style="list-style-type: none"> Nf1MyoD^{-/-}, Nf1Prx1^{-/-}, Nf1iPax7^{-/-} 	<ol style="list-style-type: none"> Summers MA, Quinlan KG, Payne JM, Little DG, North KN, Schindeler A. 2015. Skeletal muscle and motor deficits in Neurofibromatosis Type 1. <i>J Musculoskelet Neuronal Interact</i>. Jun;15(2):161-70. PMID: 26032208; PMCID: PMC5133719. Summers MA, Vasiljevski ER, Mikulec K, Peacock L, Little DG, Schindeler A. 2018. Developmental dosing with a MEK inhibitor (PDO325901) rescues myopathic features of the muscle-specific but not limb-specific Nf1 knockout mouse. <i>Mol Genet Metab</i>. Apr;123(4):518-525. doi: 10.1016/j.ymgme.2018.02.009. Epub 2018 Feb 16. PMID: 29477258. Summers MA, Rupasinghe T, Vasiljevski ER, Evesson FJ, Mikulec K, Peacock L, Quinlan KG, Cooper ST, Roessner U, Stevenson DA, Little DG, Schindeler A. 2018. Dietary intervention rescues myopathy associated with neurofibromatosis type 1. <i>Hum Mol Genet</i>. Feb 15;27(4):577-588. doi: 10.1093/hmg/ddx423. PMID: 29228356.*
NF130092	Karlyne Reilly \$606,112.00	<ul style="list-style-type: none"> CDCA7L null mice 	

*Additional references can be found in Appendix A.

Animal Models

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF150030</u>	Lei Xu \$722,625.00	<ul style="list-style-type: none"> • Orthotopic cerebellopontine angle mouse model 	<ol style="list-style-type: none"> 1. Zhao Y, Liu P, Zhang N, Chen J, Landegger LD, Wu L, Zhao F, Zhao Y, Zhang Y, Zhang J, et al. 2018. Targeting the cMET pathway augments radiation response without adverse effect on hearing in NF2 schwannoma models. <i>Proc Natl Acad Sci U S A</i>. Feb 27;115(9):E2077-E2084. doi: 10.1073/pnas.1719966115. Epub Feb 9. PMID: 29440379; PMCID: PMC5834719. 2. Chen J, Landegger LD, Sun Y, Ren J, Maimon N, Wu L, Ng MR, Chen JW, Zhang N, Zhao Y, et al. 2019. A cerebellopontine angle mouse model for the investigation of tumor biology, hearing, and neurological function in NF2-related vestibular schwannoma. <i>Nat Protoc</i>. Feb;14(2):541-555. doi: 10.1038/s41596-018-0105-7. PMID: 30617350; PMCID: PMC6571021. 3. Zhang N, Chen J, Ferraro GB, Wu L, Datta M, Jain RK, Plotkin SR, Stemmer-Rachamimov A, Xu L. 2018. Anti-VEGF treatment improves neurological function in tumors of the nervous system. <i>Exp Neurol</i>. Jan;299(Pt B):326-333. doi: 10.1016/j.expneurol.2017.09.008. Epub 2017 Sep 11. PMID: 28911884.*
<u>NF160045</u>	Lu Le \$850,500.00	<ul style="list-style-type: none"> • Novel Schwannoma mouse model 	<ol style="list-style-type: none"> 1. Chen Z, Li S, Mo J, Hawley E, Wang Y, He Y, Brosseau JP, Shipman T, Clapp DW, Carroll TJ, Le LQ. 2020. Schwannoma development is mediated by Hippo pathway dysregulation and modified by RAS/MAPK signaling. <i>JCI Insight</i>. Oct 15;5(20):e141514. doi: 10.1172/jci.insight.141514. PMID: 32960816; PMCID: PMC7605536.
<u>NF960018</u>	Nancy Ratner \$700,291.00	<ul style="list-style-type: none"> • CNPase-V12-Ha-Ras mice on an Nf1 +/- background • CNPase-V12-Ha-Ras transgenic mouse lines (3) 	<ol style="list-style-type: none"> 1. Atit RP, Mitchell K, Nguyen L, Warshawsky D, Ratner N. 2000. The neurofibromatosis type 1 (Nf1) tumor suppressor is a modifier of carcinogen-induced pigmentation and papilloma formation in C57BL/6 mice. <i>J Invest Dermatol</i>. Jun;114(6):1093-100. doi: 10.1046/j.1523-1747.2000.00994.x. PMID: 10844550; PMCID: PMC2862652. 2. Atit RP, Crowe MJ, Greenhalgh DG, Wenstrup RJ, Ratner N. 1999. The Nf1 tumor suppressor regulates mouse skin wound healing, fibroblast proliferation, and collagen deposited by fibroblasts. <i>J Invest Dermatol</i>. Jun;112(6):835-42. doi: 10.1046/j.1523-1747.1999.00609.x. PMID: 10383727; PMCID: PMC2854506. 3. Rizvi TA, Huang Y, Sidani A, Atit R, Largaespada DA, Boissy RE, Ratner N. 2002. A novel cytokine pathway suppresses glial cell melanogenesis after injury to adult nerve. <i>J Neurosci</i>. Nov 15;22(22):9831-40. doi: 10.1523/JNEUROSCI.22-22-09831.2002. PMID: 12427839; PMCID: PMC1747535.

*Additional references can be found in Appendix A.

MOUSE MODELS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF960041	Tyler Jacks \$1,186,412.00	<ul style="list-style-type: none"> Model of glioblastoma: Nf1 +/-; Trp53 +/- Nf1+/-; p53+/- cis Nf2 +/-; p53 +/- 	<ol style="list-style-type: none"> Reilly KM, Loisel DA, Bronson RT, McLaughlin ME, Jacks T. 2000. Nf1;Trp53 mutant mice develop glioblastoma with evidence of strain-specific effects. <i>Nat Genet.</i> Sep;26(1):109-13. doi: 10.1038/79075. PMID: 10973261. Cichowski K, Shih TS, Schmitt E, Santiago S, Reilly K, McLaughlin ME, Bronson RT, Jacks T. 1999. Mouse models of tumor development in neurofibromatosis type 1. <i>Science.</i> Dec 10;286(5447):2172-6. doi: 10.1126/science.286.5447.2172. PMID: 10591652. McClatchey AI, Saotome I, Mercer K, Crowley D, Gusella JF, Bronson RT, Jacks T. 1998. Mice heterozygous for a mutation at the Nf2 tumor suppressor locus develop a range of highly metastatic tumors. <i>Genes Dev.</i> Apr 15;12(8):1121-33. doi: 10.1101/gad.12.8.1121. PMID: 9553042; PMCID: PMC316711.
NF960044	Luis Parada \$1,618,665.00	Conditional knockout of Nf1: <ul style="list-style-type: none"> Nf1 flox/flox Conditional knockout of Nf1 in neurons: Synapsin 1-Cre; Nf1 flox/flox Nf1+/-; p53+/- cis 	<ol style="list-style-type: none"> Gonzalez-Zulueta M, Feldman AB, Klesse LJ, Kalb RG, Dillman JF, Parada LF, Dawson TM, Dawson VL. 2000. Requirement for nitric oxide activation of p21(ras)/extracellular regulated kinase in neuronal ischemic preconditioning. <i>Proc Natl Acad Sci U S A.</i> Jan 4;97(1):436-41. doi: 10.1073/pnas.97.1.436. PMID: 10618436; PMCID: PMC26681. Klesse LJ, Meyers KA, Marshall CJ, Parada LF. 1999. Nerve growth factor induces survival and differentiation through two distinct signaling cascades in PC12 cells. <i>Oncogene.</i> Mar 25;18(12):2055-68. doi: 10.1038/sj.onc.1202524. PMID: 10321730. Klesse LJ, Parada LF. 1998. p21 ras and phosphatidylinositol-3 kinase are required for survival of wild-type and NF1 mutant sensory neurons. <i>J Neurosci.</i> Dec 15;18(24):10420-8. doi: 10.1523/JNEUROSCI.18-24-10420.1998. PMID: 9852579; PMCID: PMC6793354.*
NF980011	Kristine Vogel \$584,552.00	<ul style="list-style-type: none"> Crossed cisNf1+/-;p53+/-mice with stratagene big blue (lacI transgenic) mice to measure mutant frequency in neural crest-derived sarcomas and to determine whether a correlation exists between neural crest phenotype and mutant frequency. 	<ol style="list-style-type: none"> Miller SJ, Li H, Rizvi TA, Huang Y, Johansson G, Bowersock J, Sidani A, Vitullo J, Vogel K, Parysek LM, DeClue JE, Ratner N. 2003. Brain lipid binding protein in axon-Schwann cell interactions and peripheral nerve tumorigenesis. <i>Mol Cell Biol.</i> Mar;23(6):2213-24. doi: 10.1128/MCB.23.6.2213-2224.2003. PMID: 12612091; PMCID: PMC149461.
NF990047	David Muir \$475,670.00	<ul style="list-style-type: none"> New mouse immunodeficient mouse strain with the Nf1 background, Nf1-+/scid. 	<ol style="list-style-type: none"> Fieber LA, González DM, Wallace MR, Muir D. 2003. Delayed rectifier K currents in NF1 Schwann cells. Pharmacological block inhibits proliferation. <i>Neurobiol Dis.</i> Jul;13(2):136-46. doi: 10.1016/s0969-9961(03)00031-7. PMID: 12828937. Muir D, Neubauer D, Lim IT, Yachnis AT, Wallace MR. 2001. Tumorigenic properties of neurofibromin-deficient neurofibroma Schwann cells. <i>Am J Pathol.</i> Feb;158(2):501-13. doi: 10.1016/S0002-9440(10)63992-2. PMID: 11159187; PMCID: PMC1850316.

*Additional references can be found in Appendix A.

Animal Models

RAT MODELS

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF050083	Allan Belzberg \$835,883.43	<ul style="list-style-type: none"> Tibial neuroma transposition model for neuroma pain and hyperalgesia 	<ol style="list-style-type: none"> Dorsi MJ, Chen L, Murinson BB, Pogatzki-Zahn EM, Meyer RA, Belzberg AJ. 2008. The tibial neuroma transposition (TNT) model of neuroma pain and hyperalgesia. <i>Pain</i>. Feb;134(3):320-334. doi: 10.1016/j.pain.2007.06.030. Epub 2007 Aug 27. PMID: 17720318.

ZEBRAFISH MODELS

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF050175	Jonathan Epstein \$1,659,063.00	<ul style="list-style-type: none"> Two zebrafish lines harboring stable germline mutations in nf1a and nf1b 	<ol style="list-style-type: none"> Lee JS, Padmanabhan A, Shin J, Zhu S, Guo F, Kanki JP, Epstein JA, Look AT. 2010. Oligodendrocyte progenitor cell numbers and migration are regulated by the zebrafish orthologs of the NF1 tumor suppressor gene. <i>Hum Mol Genet</i>. Dec 1;19(23):4643-53. doi: 10.1093/hmg/ddq395. Epub 2010 Sep 21. PMID: 20858602; PMCID: PMC3999377. Shin J, Padmanabhan A, de Groh ED, Lee JS, Haidar S, Dahlberg S, Guo F, He S, Wolman MA, Granato M, Lawson ND, Wolfe SA, Kim SH, Solnica-Krezel L, Kanki JP, Ligon KL, Epstein JA, Look AT. 2012. Zebrafish neurofibromatosis type 1 genes have redundant functions in tumorigenesis and embryonic development. <i>Dis Model Mech</i>. Nov;5(6):881-94. doi: 10.1242/dmm.009779. Epub 2012 Jul 5. PMID: 22773753; PMCID: PMC3484870. Wolman MA, de Groh ED, McBride SM, Jongens TA, Granato M, Epstein JA. 2014. Modulation of cAMP and ras signaling pathways improves distinct behavioral deficits in a zebrafish model of neurofibromatosis type 1. <i>Cell Rep</i>. Sep 11;8(5):1265-70. doi: 10.1016/j.celrep.2014.07.054. Epub 2014 Aug 28. PMID: 25176649; PMCID: PMC5850931.*
NF110108	Thomas Look \$867,846.09	<ul style="list-style-type: none"> Stable zebrafish lines expressing NF1 constructs nf1/p53/pten mutant zebrafish Zebrafish MPNST model with PDGFRA overexpression in the nf1/p53 compound mutant background Zebrafish neuroblastoma model with MYCN overexpression in the nf1 mutant background 	<ol style="list-style-type: none"> Shin J, Padmanabhan A, de Groh ED, Lee JS, Haidar S, Dahlberg S, Guo F, He S, Wolman MA, Granato M, et al. 2012. Zebrafish neurofibromatosis type 1 genes have redundant functions in tumorigenesis and embryonic development. <i>Dis Model Mech</i>. Nov;5(6):881-94. doi: 10.1242/dmm.009779. Epub 2012 Jul 5. PMID: 22773753; PMCID: PMC3484870. Wolman MA, de Groh ED, McBride SM, Jongens TA, Granato M, Epstein JA. 2014. Modulation of cAMP and ras signaling pathways improves distinct behavioral deficits in a zebrafish model of neurofibromatosis type 1. <i>Cell Rep</i>. Sep 11;8(5):1265-70. doi: 10.1016/j.celrep.2014.07.054. Epub 2014 Aug 28. PMID: 25176649; PMCID: PMC5850931. Zhu C, Smith T, McNulty J, Rayla AL, Lakshmanan A, Siekmann AF, Buffardi M, Meng X, Shin J, Padmanabhan A, et al. 2011. Evaluation and application of modularly assembled zinc-finger nucleases in zebrafish. <i>Development</i>. Oct;138(20):4555-64. doi: 10.1242/dev.066779. PMID: 21937602; PMCID: PMC3177320.*
NF130052	Michael Granato	<ul style="list-style-type: none"> nf1a+/-; nf1b-/- exhibit memory and learning deficits 	<ol style="list-style-type: none"> Wolman MA, de Groh ED, McBride SM, Jongens TA, Granato M, Epstein JA. 2014. Modulation of cAMP and ras signaling pathways improves distinct behavioral deficits in a zebrafish model of neurofibromatosis type 1. <i>Cell Rep</i>. Sep 11;8(5):1265-70. doi: 10.1016/j.celrep.2014.07.054. Epub 2014 Aug 28. PMID: 25176649; PMCID: PMC5850931.

*Additional references can be found in Appendix A.

Antibodies

Antibodies are blood proteins generated within the immune system that recognize and nullify foreign substances in the body such as bacteria and viruses. Antibodies possess high specificity and have become very useful in many laboratory techniques which aid in detection, diagnosis and measurement of biological markers. They bind easily to other substances and allow for easier identification of molecules that cannot otherwise be seen by the natural eye. Both monoclonal antibodies (derived from a homogenous mix of immune cells and binding to the same epitope of an antigen) and polyclonal antibodies (derived from a heterogeneous mix of immune cells and binding to the different epitopes of an antigen) are utilized in NF research.

ANTIBODIES

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF000002</u>	Dennis LaJeunesse \$371,050.00	<ul style="list-style-type: none"> • Polyclonal antibodies that recognize Drosophila scribbler and human homologue to scribbler (HSbb) 	<ol style="list-style-type: none"> 1. LaJeunesse DR, Buckner SM, Lake J, Na C, Pirt A, Fromson K. 2004. Three new Drosophila markers of intracellular membranes. <i>Biotechniques</i>. May;36(5):784-8, 790. doi: 10.2144/04365ST01. PMID: 15152597. 2. LaJeunesse D. 2010. Alteration of wing size through over-expression of scribbler isoforms. <i>Fly (Austin)</i>. Oct-Dec;4(4):320-6. doi: 10.4161/fly.4.4.13148. Epub Oct 1. PMID: 20798603.
<u>NF010016</u>	Marco Giovannini and David Gutmann \$785,230.00	<ul style="list-style-type: none"> • Polyclonal rabbit anti-TSLC1 and polyclonal rabbit anti-protein 4.1B 	<ol style="list-style-type: none"> 1. Surace EI, Lusia E, Murakami Y, Scheithauer BW, Perry A, Gutmann DH. 2004. Loss of tumor suppressor in lung cancer-1 (TSLC1) expression in meningioma correlates with increased malignancy grade and reduced patient survival. <i>J Neuropathol Exp Neurol</i>. Oct;63(10):1015-27. doi: 10.1093/jnen/63.10.1015. PMID: 15535129. 2. McClatchey AI, Giovannini M. 2005. Membrane organization and tumorigenesis--the NF2 tumor suppressor, Merlin. <i>Genes Dev</i>. Oct 1;19(19):2265-77. doi: 10.1101/gad.1335605. PMID: 16204178.
<u>NF020037</u>	Wallace Ip \$690,750.00	<ul style="list-style-type: none"> • Two monoclonal antibodies against merlin. • Two monoclonal antibodies and two polyclonal antibodies against merlin. • Two monoclonal antibodies against tropomodulin3. • Two monoclonal antibodies against PPM1B2. 	<ol style="list-style-type: none"> 1. Stickney JT, Bacon WC, Rojas M, Ratner N, Ip W. 2004. Activation of the tumor suppressor merlin modulates its interaction with lipid rafts. <i>Cancer Res</i>. Apr 15;64(8):2717-24. doi: 10.1158/0008-5472.can-03-3798. PMID: 15087385. 2. Meng JJ, Rojas M, Bacon W, Stickney JT, Ip W. 2005. Methods to study protein-protein interactions. <i>Methods Mol Biol</i>. 289:341-58. doi: 10.1385/1-59259-830-7:341. PMID: 15502197. 3. Kumar G, Meng JJ, Ip W, Co CC, Ho CC. 2005. Cell motility assays on tissue culture dishes via non-invasive confinement and release of cells. <i>Langmuir</i>. Sep 27;21(20):9267-73. doi: 10.1021/la050332n. PMID: 16171361.
<u>NF020087</u>	Athar Chishti \$216,000.00	<ul style="list-style-type: none"> • Human p55 monoclonal antibody (2G4, IgG2b) 	<ol style="list-style-type: none"> 1. Seo PS, Quinn BJ, Khan AA, Zeng L, Takoudis CG, Hanada T, Bolis A, Bolino A, Chishti AH. 2009. Identification of erythrocyte p55/MPP1 as a binding partner of NF2 tumor suppressor protein/Merlin. <i>Exp Biol Med (Maywood)</i>. Mar;234(3):255-62. doi: 10.3181/0809-RM-275. Epub 2009 Jan 14. PMID: 19144871; PMCID: PMC3959803.

Antibodies

ANTIBODIES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF030020</u>	David Gutmann \$1,183,129.00	<ul style="list-style-type: none"> Polyclonal rabbit anti-protein 4.1B 	<ol style="list-style-type: none"> Robb VA, Gerber MA, Hart-Mahon EK, Gutmann DH. 2005. Membrane localization of the U2 domain of Protein 4.1B is necessary and sufficient for meningioma growth suppression. <i>Oncogene</i>. Mar 10;24(11):1946-57. doi: 10.1038/sj.onc.1208335. PMID: 15688033. Singh V, Miranda TB, Jiang W, Frankel A, Roemer ME, Robb VA, Gutmann DH, Herschman HR, Clarke S, Newsham IF. 2004. 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. <i>Oncogene</i>. Oct 14;23(47):7761-71. doi: 10.1038/sj.onc.1208057. PMID: 15334060. 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. 2006. Promoter hypermethylation of the potential tumor suppressor DAL-1/4.1B gene in renal clear cell carcinoma. <i>Int J Cancer</i>. Feb 15;118(4):916-23. doi: 10.1002/ijc.21450. PMID: 16152585.*
<u>NF043043</u>	Wallace Ip \$1,446,167.00	<ul style="list-style-type: none"> Monoclonal and polyclonal merlin antibodies Monoclonal and polyclonal PPM1B2 antibodies 	<ol style="list-style-type: none"> Lepont P, Stickney JT, Foster LA, Meng JJ, Hennigan RF, Ip W. 2008. Point mutation in the NF2 gene of HEI-193 human schwannoma cells results in the expression of a merlin isoform with attenuated growth suppressive activity. <i>Mutat Res</i>. Jan 1;637(1-2):142-51. doi: 10.1016/j.mrfmmm.2007.07.015. Epub 2007 Aug 6. PMID: 17868749; PMCID: PMC2233940. Hennigan RF, Chaiken MF, Foster LA, Ip W. 2009. A FRET-based approach for studying conformational changes of a cytoskeleton-related tumor suppressor molecule. <i>Methods Mol Biol</i>. 586:143-56. doi: 10.1007/978-1-60761-376-3_7. PMID: 19768428. Hennigan RF, Foster LA, Chaiken MF, Mani T, Gomes MM, Herr AB, Ip W. 2010. Fluorescence resonance energy transfer analysis of merlin conformational changes. <i>Mol Cell Biol</i>. Jan;30(1):54-67. doi: 10.1128/MCB.00248-09. PMID: 19884346; PMCID: PMC2798298.*
<u>NF960027</u>	Margaret Wallace \$594,663.00	<ul style="list-style-type: none"> Polyclonal rabbit anti-neurofibromin: PcNFn27 Monoclonal rabbit anti-neurofibromin: McNFn27a and McNFn27b 	<ol style="list-style-type: none"> Muir D, Neubauer D, Lim IT, Yachnis AT, Wallace MR. 2001. Tumorigenic properties of neurofibromin-deficient neurofibroma Schwann cells. <i>Am J Pathol</i>. Feb;158(2):501-13. doi: 10.1016/S0002-9440(10)63992-2. PMID: 11159187; PMCID: PMC1850316.

*Additional references can be found in Appendix A.

Cell and Molecular Bio Tools

These involve techniques that study the structure and function of cells and other macromolecules such as DNA, RNA, lipids and proteins. The study of biological activity at a cellular and molecular level in NF is useful for sorting and/or isolating such distinct entities in order to enhance their understanding.

CELL AND MOLECULAR BIO TOOLS

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF000002	Dennis LaJeunesse \$371,050.00	<ul style="list-style-type: none"> RNA probes specific to SbbA, SbbB, and HSbb FLAG-tagged versions of scribbler (Sbb) Flag and HA-tagged HSbb transgenes for examination in human tissue cell cultures 	<ol style="list-style-type: none"> LaJeunesse DR, Buckner SM, Lake J, Na C, Pirt A, Fromson K. 2004. Three new Drosophila markers of intracellular membranes. <i>Biotechniques</i>. May;36(5):784-8, 790. doi: 10.2144/04365ST01. PMID: 15152597. LaJeunesse D. 2010. Alteration of wing size through over-expression of scribbler isoforms. <i>Fly (Austin)</i>. Oct-Dec;4(4):320-6. doi: 10.4161/fly.4.4.13148. Epub 2010 Oct 1. PMID: 20798603.
NF010022	Qin Yu \$475,500.00	<ul style="list-style-type: none"> Constructs for deletion mutants of merlin 	<ol style="list-style-type: none"> Bai Y, Liu YJ, Wang H, Xu Y, Stamenkovic I, Yu Q. 2007. Inhibition of the hyaluronan-CD44 interaction by merlin contributes to the tumor-suppressor activity of merlin. <i>Oncogene</i>. Feb 8;26(6):836-50. doi: 10.1038/sj.onc.1209849. Epub 2006 Sep 4. PMID: 16953231. Xu Y, Yu Q. 2003. E-cadherin negatively regulates CD44-hyaluronan interaction and CD44-mediated tumor invasion and branching morphogenesis. <i>J Biol Chem</i>. Mar 7;278(10):8661-8. doi: 10.1074/jbc.M208181200. Epub 2003 Jan 2. PMID: 12511569.
NF010126	Stephen Lambert \$475,876.00	<ul style="list-style-type: none"> PI has developed a myelinating explant system in order to study the transition of Schwann cell precursors to the differentiated bipolar stage. 	<ol style="list-style-type: none"> Gatto CL, Walker BJ, Lambert S. 2007. Asymmetric ERM activation at the Schwann cell process tip is required in axon-associated motility. <i>J Cell Physiol</i>. Jan;210(1):122-32. doi: 10.1002/jcp.20844. PMID: 17061246.
NF020008	Matt Van de Rijn	<p>mRNA and miRNA expression profiles for human MPNSTs, neurofibromas, schwannomas, and synovial sarcomas</p> <ul style="list-style-type: none"> A new tissue array (TA-138) has been constructed to include 68 MPNSTs, 42 NFs, 22 SWs, and 15 SSs. In situ hybridization probes have been generated for 14 genes (identified under key research accomplishments). Tissue microarray of nerve sheath tumors miRNA microarrays containing 668 probes small RNA libraries of 4 sarcomas (1 MPNST, 1NF and 2 SS) tissues 14 overrepresented and 5 underrepresented miRNAs in MPNSTs 	<ol style="list-style-type: none"> Terry J, Saito T, Subramanian S, Ruttan C, Antonescu CR, Goldblum JR, Downs-Kelly E, Corless CL, Rubin BP, van de Rijn M, Ladanyi M, Nielsen TO. 2007. TLE1 as a diagnostic immunohistochemical marker for synovial sarcoma emerging from gene expression profiling studies. <i>Am J Surg Pathol</i>. Feb;31(2):240-6. doi: 10.1097/01.pas.0000213330.71745.39. PMID: 17255769. Subramanian S, Thayanithy V, West RB, Lee CH, Beck AH, Zhu S, Downs-Kelly E, Montgomery K, Goldblum JR, Hogendoorn PC, Corless CL, Oliveira AM, Dry SM, Nielsen TO, Rubin BP, Fletcher JA, Fletcher CD, van de Rijn M. 2010. Genome-wide transcriptome analyses reveal p53 inactivation mediated loss of miR-34a expression in malignant peripheral nerve sheath tumours. <i>J Pathol</i>. Jan;220(1):58-70. doi: 10.1002/path.2633. PMID: 19890883; PMCID: PMC4058327.

Cell and Molecular Bio Tools

CELL AND MOLECULAR BIO TOOLS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF020061	Stefan Mundlos \$405,172.00	<ul style="list-style-type: none"> A plasmid was generated for control of Cre by the osteoclast-specific tartrate-resistant acid phosphatase (TRAP) promoter. 	
NF020087	Athar Chishti \$216,000.00	<ul style="list-style-type: none"> Human GST- p55(SH3 and GUK domain) fusion construct Maltose-binding protein fusions of recombinant NF2 N and C-terminus construct Wild type and mutant recombinant p55 full-length construct Recombinant protein 4.1 construct 	<ol style="list-style-type: none"> Seo PS, Quinn BJ, Khan AA, Zeng L, Takoudis CG, Hanada T, Bolis A, Bolino A, Chishti AH. 2009. Identification of erythrocyte p55/ MPP1 as a binding partner of NF2 tumor suppressor protein/ Merlin. <i>Exp Biol Med</i> (Maywood). Mar;234(3):255-62. doi: 10.3181/0809-RM-275. Epub 2009 Jan 14. PMID: 19144871; PMCID: PMC3959803. Jindal HK, Yoshinaga K, Seo PS, Lutchman M, Dion PA, Rouleau GA, Hanada T, Chishti AH. 2006. Purification of the NF2 tumor suppressor protein from human erythrocytes. <i>Can J Neurol Sci</i>. Nov;33(4):394-402. doi: 10.1017/s0317167100005357. PMID: 17168165.
NF030063	Shao-Jun Tang \$666,174.31	<ul style="list-style-type: none"> &#945;CaMKII and CREB1 Lovastatin Genes involved in the regulation of synaptic transmission, synaptic plasticity, and memory formation. 	<ol style="list-style-type: none"> Park CS, Zhong L, Tang SJ. 2009. Aberrant expression of synaptic plasticity-related genes in the NF1+/- mouse hippocampus. <i>J Neurosci Res</i>. Nov 1;87(14):3107-19. doi: 10.1002/jnr.22134. PMID: 19475561.
NF030070	Long-Sheng Chang \$636,452.00	<ul style="list-style-type: none"> pUASP myc-tagged merlin constructs: Mer+ (wild type) Mer3 mutant (Met177Ile) MerΔBB mutant (Blue Box deletion) Mer1-379 mutant Mer345-635 mutant 	<ol style="list-style-type: none"> Golovnina K, Blinov A, Akhmametyeva EM, Omelyanchuk LV, Chang LS. 2005. Evolution and origin of merlin, the product of the Neurofibromatosis type 2 (NF2) tumor-suppressor gene. <i>BMC Evol Biol</i>. Dec 2;5:69. doi: 10.1186/1471-2148-5-69. PMID: 16324214; PMCID: PMC1315344. Neff BA, Welling DB, Akhmametyeva E, Chang LS. 2006. The molecular biology of vestibular schwannomas: dissecting the pathogenic process at the molecular level. <i>Otol Neurotol</i>. Feb;27(2):197-208. doi: 10.1097/01.mao.0000180484.24242.54. PMID: 16436990. Neff BA, Oberstien E, Lorenz M, Chaudhury AR, Welling DB, Chang LS. 2006. Cyclin D(1) and D(3) expression in vestibular schwannomas. <i>Laryngoscope</i>. Mar;116(3):423-6. doi: 10.1097/01.mlg.0000195076.05466.6c. PMID: 16540902.*

*Additional references can be found in Appendix A.

CELL AND MOLECULAR BIO TOOLS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF043008	Olli Carpen \$630,000.00	<ul style="list-style-type: none"> • Retroviral plasmid constructs of wild-type ezrin and ezrin Y477F, ezrin Y145F, and ezrin Y353F mutants for expression in mammalian cells. • Plasmid constructs with altered merlin S10 and S518 residues. • merlin's N-terminus phosphospecific antibody • merlin Bimolecular fluorescence complementation vector constructs • ezrin Bimolecular fluorescence complementation vector constructs • merlin S10A and S518D double mutant construct • merlin E545K and E547K mutation constructs 	<ol style="list-style-type: none"> 1. Grönholm M, Muranen T, Toby GG, Utermark T, Hanemann CO, Golemis EA, Carpén O. 2006. A functional association between merlin and HEI10, a cell cycle regulator. <i>Oncogene</i>. Jul 27;25(32):4389-98. doi: 10.1038/sj.onc.1209475. Epub Mar 13. PMID: 16532029. 2. Heiska L, Carpén O. 2005. Src phosphorylates ezrin at tyrosine 477 and induces a phosphospecific association between ezrin and a kelch-repeat protein family member. <i>J Biol Chem</i>. Mar 18;280(11):10244-52. doi: 10.1074/jbc.M411353200. Epub 2004 Dec 28. PMID: 15623525. 3. Muranen T, Grönholm M, Renkema GH, Carpén O. 2005. Cell cycle-dependent nucleocytoplasmic shuttling of the neurofibromatosis 2 tumour suppressor merlin. <i>Oncogene</i>. Feb 10;24(7):1150-8. doi: 10.1038/sj.onc.1208283. PMID: 15580288.*
NF043040	Andrea McClatchey \$830,034.00	<ul style="list-style-type: none"> • PDZ domain-containing adaptor NHE-RF • Merlin:NHE-RF:EGFR:E-cadherin complex • NHE-RF1 	<ol style="list-style-type: none"> 1. Curto M, Cole BK, Lallemand D, Liu CH, McClatchey AI. 2007. Contact-dependent inhibition of EGFR signaling by Nf2/Merlin. <i>J Cell Biol</i>. Jun 4;177(5):893-903. doi: 10.1083/jcb.200703010. PMID: 17548515; PMCID: PMC2064288. 2. Cole BK, Curto M, Chan AW, McClatchey AI. 2008. Localization to the cortical cytoskeleton is necessary for Nf2/merlin-dependent epidermal growth factor receptor silencing. <i>Mol Cell Biol</i>. Feb;28(4):1274-84. doi: 10.1128/MCB.01139-07. Epub 2007 Dec 17. PMID: 18086884; PMCID: PMC2258752. 3. Curto M, McClatchey AI. 2008. Nf2/Merlin: a coordinator of receptor signalling and intercellular contact. <i>Br J Cancer</i>. Jan 29;98(2):256-62. doi: 10.1038/sj.bjc.6604002. Epub 2007 Oct 30. PMID: 17971776; PMCID: PMC2361439.*
NF043043	Wallace Ip \$1,446,167.00	<ul style="list-style-type: none"> • Adenoviral and lentiviral merlin and PPM1B constructs • FRET probes of merlin conformation – C-merlin-V, C-merlin, merlin-V, C6V, C-merlinN-half, merlinC-half-V, in mammalian expression vector • Deletion constructs of merlin for FRET – C-FERM-V, C-FERM-Helix-V, C-helix-CTD-V • FRET probes of merlin conformation in bacterial expression vector • Wildtype GST-PAK1 fusion protein • PPM1B2 mutants D60A, D243A, and R179G • MYPT1-PP1 • CPI-17 	<ol style="list-style-type: none"> 1. Lepont P, Stickney JT, Foster LA, Meng JJ, Hennigan RF, Ip W. 2008. Point mutation in the NF2 gene of HEI-193 human schwannoma cells results in the expression of a merlin isoform with attenuated growth suppressive activity. <i>Mutat Res</i>. Jan 1;637(1-2):142-51. doi: 10.1016/j.mrfmmm.2007.07.015. Epub 2007 Aug 6. PMID: 17868749; PMCID: PMC2233940. 2. Hennigan RF, Chaiken MF, Foster LA, Ip W. 2009. A FRET-based approach for studying conformational changes of a cytoskeleton-related tumor suppressor molecule. <i>Methods Mol Biol</i>. 586:143-56. doi: 10.1007/978-1-60761-376-3_7. PMID: 19768428. 3. Hennigan RF, Foster LA, Chaiken MF, Mani T, Gomes MM, Herr AB, Ip W. 2010. Fluorescence resonance energy transfer analysis of merlin conformational changes. <i>Mol Cell Biol</i>. Jan;30(1):54-67. doi: 10.1128/MCB.00248-09. PMID: 19884346; PMCID: PMC2798298.*
NF043064	Karen Cichowski \$799,983.00	<ul style="list-style-type: none"> • NF1 mammalian expression vector 	

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Cell and Molecular Bio Tools

CELL AND MOLECULAR BIO TOOLS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF050009	Keqiang Ye \$1,110,363.00	<ul style="list-style-type: none"> S315A, T230A, T230AS315A, S315D, T230D, T230DS315D mutants in the context of the full-length merlin molecule - constructs 	<ol style="list-style-type: none"> Tang X, Jang SW, Wang X, Liu Z, Bahr SM, Sun SY, Brat D, Gutmann DH, Ye K. 2007. Akt phosphorylation regulates the tumour-suppressor merlin through ubiquitination and degradation. <i>Nat Cell Biol.</i> Oct;9(10):1199-207. doi: 10.1038/ncb1641. Epub 2007 Sep 23. PMID: 17891137. Okada M, Wang Y, Jang SW, Tang X, Neri LM, Ye K. 2009. Akt phosphorylation of merlin enhances its binding to phosphatidylinositols and inhibits the tumor-suppressive activities of merlin. <i>Cancer Res.</i> May 1;69(9):4043-51. doi: 10.1158/0008-5472.CAN-08-3931. Epub 2009 Apr 7. PMID: 19351837; PMCID: PMC2810124.
NF050130	Jason Weber \$99,950.31	<ul style="list-style-type: none"> Dominant negative Rac1 mutant (Rac1N17) construct S6K1 construct Dominant negative NPM mutant (NPMdL) construct 	<ol style="list-style-type: none"> Sandsmark DK, Pelletier C, Weber JD, Gutmann DH. 2007. Mammalian target of rapamycin: master regulator of cell growth in the nervous system. <i>Histol Histopathol.</i> Aug;22(8):895-903. doi: 10.14670/HH-22.895. PMID: 17503347.
NF073146	Susan Lindquist \$1,023,750.00	<ul style="list-style-type: none"> pBabe vector encoding EGFP and pBabe vector encoding dominant-negative MEK1 mutant (Ser218Ala and Ser222Ala) 	<ol style="list-style-type: none"> Dai C, Santagata S, Tang Z, Shi J, Cao J, Kwon H, Bronson RT, Whitesell L, Lindquist S. 2012. Loss of tumor suppressor NF1 activates HSF1 to promote carcinogenesis. <i>J Clin Invest.</i> Oct;122(10):3742-54. doi: 10.1172/JCI62727. Epub 2012 Sep 4. PMID: 22945628; PMCID: PMC3461912. Whitesell L, Lindquist S. 2009. Inhibiting the transcription factor HSF1 as an anticancer strategy. <i>Expert Opin Ther Targets.</i> Apr;13(4):469-78. doi: 10.1517/14728220902832697. PMID: 19335068.
NF080062	Lawrence Kirschner \$776,420.14	<ul style="list-style-type: none"> pcDNA3.1 encoding wildtype Rac1, Rac1 Q61L, or Rac1 T17N 	<ol style="list-style-type: none"> Manchanda PK, Jones GN, Lee AA, Pringle DR, Zhang M, Yu L, La Perle KM, Kirschner LS. 2013. Rac1 is required for Prkar1a-mediated Nf2 suppression in Schwann cell tumors. <i>Oncogene.</i> Jul 25;32(30):3491-9. doi: 10.1038/onc.2012.374. Epub 2012 Oct 8. PMID: 23045281; PMCID: PMC3542412. Yin Z, Pringle DR, Jones GN, Kelly KM, Kirschner LS. 2011. Differential role of PKA catalytic subunits in mediating phenotypes caused by knockout of the Carney complex gene Prkar1a. <i>Mol Endocrinol.</i> Oct;25(10):1786-93. doi: 10.1210/me.2011-1008. Epub 2011 Aug 18. PMID: 21852354; PMCID: PMC3182421.
NF093039	Jonathan Chernoff \$174,348.00	<ul style="list-style-type: none"> Nf1 retargeting vector to repair NF1 allele in pluripotent stem cells NF1 gene repair vector designed to replace the disrupted Nf1 allele in a mouse model by restoring exon 31 	
NF093079	Pamela Vanderzalm \$156,000.00	<ul style="list-style-type: none"> Kinase domains of Hpo (aa. 1-367), MST1 (aa 1-320), and MST2(aa 1-316) in pGEX expression vector 	<ol style="list-style-type: none"> Boggiano JC, Vanderzalm PJ, Fehon RG. 2011. Tao-1 phosphorylates Hippo/MST kinases to regulate the Hippo-Salvador-Warts tumor suppressor pathway. <i>Dev Cell.</i> Nov 15;21(5):888-95. doi: 10.1016/j.devcel.2011.08.028. PMID: 22075147; PMCID: PMC3217187.

CELL AND MOLECULAR BIO TOOLS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF100160	Toshifumi Tomoda \$664,000.00	<ul style="list-style-type: none"> Plasmids for the autophagy-related protein Unc51.Atg1-myc and the NF2 point mutation Merlin K79E-myc 	<ol style="list-style-type: none"> Flores R (Third), Hirota Y, Armstrong B, Sawa A, Tomoda T. 2011. DISC1 regulates synaptic vesicle transport via a lithium-sensitive pathway. <i>Neurosci Res. Sep;71(1):71-7</i>. doi: 10.1016/j.neures.2011.05.014. Epub 2011 Jun 12. PMID: 21664390; PMCID: PMC3156137.

Cell Lines

These are groups of cells from a multicellular organism that continue to undergo proliferation due to mutation(s) and as such can be grown for research purposes for extended periods of time. NF-specific cells generated from patients can provide a stable repository to be used for testing the safety and effectiveness of different therapies

CELL LINES

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF010022	Qin Yu \$475,500.00	<ul style="list-style-type: none"> Tr6BC1 and TA3 cell lines expressing wild type merlin or different deletion mutants of merlin 	<ol style="list-style-type: none"> Bai Y, Liu YJ, Wang H, Xu Y, Stamenkovic I, Yu Q. 2007. Inhibition of the hyaluronan-CD44 interaction by merlin contributes to the tumor-suppressor activity of merlin. <i>Oncogene</i>. Feb 8;26(6):836-50. doi: 10.1038/sj.onc.1209849. Epub 2006 Sep 4. PMID: 16953231. Xu Y, Yu Q. 2003. E-cadherin negatively regulates CD44-hyaluronan interaction and CD44-mediated tumor invasion and branching morphogenesis. <i>J Biol Chem</i>. Mar 7;278(10):8661-8. doi: 10.1074/jbc.M208181200. Epub 2003 Jan 2. PMID: 12511569.
NF020037	Wallace Ip \$690,750.00	<ul style="list-style-type: none"> NIH3T3 and COS-7 lines stably expressing various mutant merlins. 	<ol style="list-style-type: none"> Stickney JT, Bacon WC, Rojas M, Ratner N, Ip W. 2004. Activation of the tumor suppressor merlin modulates its interaction with lipid rafts. <i>Cancer Res</i>. Apr 15;64(8):2717-24. doi: 10.1158/0008-5472.can-03-3798. PMID: 15087385. Meng JJ, Rojas M, Bacon W, Stickney JT, Ip W. 2005. Methods to study protein-protein interactions. <i>Methods Mol Biol</i>. 289:341-58. doi: 10.1385/1-59259-830-7:341. PMID: 15502197.
NF030039	Robert Martuza \$2,647,746.00	<ul style="list-style-type: none"> Mouse schwannoma cell line Nf2S-1 	<ol style="list-style-type: none"> Messerli SM, Prabhakar S, Tang Y, Mahmood U, Giovannini M, Weissleder R, Bronson R, Martuza R, Rabkin S, Breakefield XO. 2006. Treatment of schwannomas with an oncolytic recombinant herpes simplex virus in murine models of neurofibromatosis type 2. <i>Hum Gene Ther</i>. Jan;17(1):20-30. doi: 10.1089/hum.2006.17.20. Erratum in: <i>Hum Gene Ther</i>. Jul;17(7):796. Giovannini, Marco [added]. PMID: 16409122. Prabhakar S, Messerli S, Rabkin S, Martuza R, Breakefield XO. Treatment of experimental schwannomas with HSV recombinant and amplicon vectors. 2006. <i>Molecular Therapy</i>. May;Milwaukee 13(S1):S59. DOI:10.1016/j.ymthe.2006.08.174. Liu TC, Zhang T, Fukuhara H, Kuroda T, Todo T, Canron X, Bikfalvi A, Martuza RL, Kurtz A, Rabkin SD. 2006. Dominant-negative fibroblast growth factor receptor expression enhances antitumoral potency of oncolytic herpes simplex virus in neural tumors. <i>Clin Cancer Res</i>. Nov 15;12(22):6791-9. doi: 10.1158/1078-0432.CCR-06-0263. PMID: 17121900.*
NF043040	Andrea McClatchey \$830,034.00	<ul style="list-style-type: none"> Nf2frtSTOPfrt/lox cells Nf2-/- cells 	<ol style="list-style-type: none"> Curto M, Cole BK, Lallemand D, Liu CH, McClatchey AI. 2007. Contact-dependent inhibition of EGFR signaling by Nf2/Merlin. <i>J Cell Biol</i>. Jun 4;177(5):893-903. doi: 10.1083/jcb.200703010. PMID: 17548515; PMCID: PMC2064288. Cole BK, Curto M, Chan AW, McClatchey AI. 2008. Localization to the cortical cytoskeleton is necessary for Nf2/merlin-dependent epidermal growth factor receptor silencing. <i>Mol Cell Biol</i>. Feb;28(4):1274-84. doi: 10.1128/MCB.01139-07. Epub 2007 Dec 17. PMID: 18086884; PMCID: PMC2258752. Curto M, McClatchey AI. 2008. Nf2/Merlin: a coordinator of receptor signalling and intercellular contact. <i>Br J Cancer</i>. Jan 29;98(2):256-62. doi: 10.1038/sj.bjc.6604002. Epub 2007 Oct 30. PMID: 17971776; PMCID: PMC2361439.*

*Additional references can be found in Appendix A.

CELL LINES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF043043	Wallace Ip \$1,446,167.00	<ul style="list-style-type: none"> Immortalized mouse embryo fibroblasts derived from Nf2^{-/-} mouse NIH3T3, Cos7 and HEI-193 lines expressing various mutant merlins 	<ol style="list-style-type: none"> Lepont P, Stickney JT, Foster LA, Meng JJ, Hennigan RF, Ip W. 2008. Point mutation in the NF2 gene of HEI-193 human schwannoma cells results in the expression of a merlin isoform with attenuated growth suppressive activity. <i>Mutat Res.</i> Jan 1;637(1-2):142-51. doi: 10.1016/j.mrfmmm.2007.07.015. Epub 2007 Aug 6. PMID: 17868749; PMCID: PMC2233940.
NF050009	Keqiang Ye \$1,110,363.00	<ul style="list-style-type: none"> purified GST-merlin-NTD. GST-merlin Merlin S315. Merlin-expressing RT4-D6P2T rat schwannoma cells. HEK 293 cells were transfected with wildtype and S315A mutant merlin constructs RT4-D6P2T rat schwannoma cells with doxycycline-regulatable wild-type (54 and 67) or (L64P) mutant merlin. HEK293 cells transfected with myc-PIKE-L. RT4 rat schwannoma cell lines with nonphosphorylatable (T230AS315A) and phosphorylation-mimetic (T230DS315D)merlin mutants. HEK293 transfected with HA-merlin. HEK293 transfected with HA-ubiquitin. 	<ol style="list-style-type: none"> Tang X, Jang SW, Wang X, Liu Z, Bahr SM, Sun SY, Brat D, Gutmann DH, Ye K. 2007. Akt phosphorylation regulates the tumour-suppressor merlin through ubiquitination and degradation. <i>Nat Cell Biol.</i> Oct;9(10):1199-207. doi: 10.1038/ncb1641. Epub 2007 Sep 23. PMID: 17891137. Okada M, Wang Y, Jang SW, Tang X, Neri LM, Ye K. 2009. Akt phosphorylation of merlin enhances its binding to phosphatidylinositols and inhibits the tumor-suppressive activities of merlin. <i>Cancer Res.</i> May 1;69(9):4043-51. doi: 10.1158/0008-5472.CAN-08-3931. Epub Apr 7. PMID: 19351837; PMCID: PMC2810124.
NF050061	Anita Lal \$668,861.06	<ul style="list-style-type: none"> Immortalized arachnoidal (AC1) and meningioma (MENII-1) cell lines with stable NF2 suppression which can be used for xenografts KT21MG1-Merlin SA2-ControlsiRNA1 SA2- NF2siRNA3 SF6717-ControlsiRNA1 SF6717-NF2siRNA3 Developed meningioma-specific in vivo Neurofibromatosis 2 model systems. 	<ol style="list-style-type: none"> Striedinger K, VandenBerg SR, Baia GS, McDermott MW, Gutmann DH, Lal A. 2008. The neurofibromatosis 2 tumor suppressor gene product, merlin, regulates human meningioma cell growth by signaling through YAP. <i>Neoplasia.</i> Nov;10(11):1204-12. doi: 10.1593/neo.08642. PMID: 18953429; PMCID: PMC2570596.

Cell Lines

CELL LINES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF050145	Victor-Felix Mautner \$628,550.00	<p>MPNST cell lines:</p> <ul style="list-style-type: none"> • 1507 (loss of heterozygosity in p53; splicing mutation in intron 23-1 and a deletion in exon 10a of NF1) • 1844 (loss of heterozygosity in NF1 region) 	<ol style="list-style-type: none"> 1. Demestre M, Messerli SM, Celli N, Shahhossini M, Kluwe L, Mautner V, Maruta H. 2009. CAPE (caffeic acid phenethyl ester)-based propolis extract (Bio 30) suppresses the growth of human neurofibromatosis (NF) tumor xenografts in mice. <i>Phytother Res.</i> Feb;23(2):226-30. doi: 10.1002/ptr.2594. PMID: 18726924. 2. Holtkamp N, Malzer E, Zietsch J, Okuducu AF, Mucha J, Mawrin C, Mautner VF, Schildhaus HU, von Deimling A. 2008. EGFR and erbB2 in malignant peripheral nerve sheath tumors and implications for targeted therapy. <i>Neuro Oncol.</i> Dec;10(6):946-57. doi: 10.1215/15228517-2008-053. Epub 2008 Jul 23. PMID: 18650488; PMCID: PMC2719009. 3. Holtkamp N, Okuducu AF, Mucha J, Afanasieva A, Hartmann C, Atallah I, Estevez-Schwarz L, Mawrin C, Friedrich RE, Mautner VF, von Deimling A. 2006. Mutation and expression of PDGFRA and KIT in malignant peripheral nerve sheath tumors, and its implications for imatinib sensitivity. <i>Carcinogenesis.</i> Mar;27(3):664-71. doi: 10.1093/carcin/bgj273. Epub 2005 Dec 15. PMID: 16357008.*
NF060049	Samuel Rabkin \$2,193,808.00	<ul style="list-style-type: none"> • Human MPNST stem-like cell (MSLC) line S462 MSLCs 	<ol style="list-style-type: none"> 1. Demestre M, Herzberg J, Holtkamp N, Hagel C, Reuss D, Friedrich RE, Kluwe L, Von Deimling A, Mautner VF, Kurtz A. 2010. Imatinib mesylate (Glivec) inhibits Schwann cell viability and reduces the size of human plexiform neurofibroma in a xenograft model. <i>J Neurooncol.</i> May;98(1):11-9. doi: 10.1007/s11060-009-0049-4. Epub 2009 Nov 17. PMID: 19921098. 2. Kanai R, Wakimoto H, Cheema T, Rabkin SD. 2010. Oncolytic herpes simplex virus vectors and chemotherapy: are combinatorial strategies more effective for cancer? <i>Future Oncol.</i> Apr;6(4):619-34. doi: 10.2217/fon.10.18. PMID: 20373873; PMCID: PMC2904234. 3. Spyra M, Kluwe L, Hagel C, Nguyen R, Panse J, Kurtz A, Mautner VF, Rabkin SD, Demestre M. 2011. Cancer stem cell-like cells derived from malignant peripheral nerve sheath tumors. <i>PLoS One.</i> 6(6):e21099. doi: 10.1371/journal.pone.0021099. Epub 2011 Jun 13. PMID: 21695156; PMCID: PMC3113907.*
NF093039	Jonathan Chernoff \$174,348.00	<ul style="list-style-type: none"> • Nf1+/- iPS cell lines 	

*Additional references can be found in Appendix A.

CELL LINES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF110082	Steven Carroll \$769,125.00	<ul style="list-style-type: none"> • Early passage cultures from 90 P0-GGFβ3 MPNSTs • Early passage cultures from 12 P0-GGFβ3;Trp53+/- MPNSTs 	<ol style="list-style-type: none"> 1. Kazmi SJ, Byer SJ, Eckert JM, Turk AN, Huijbregts RP, Brossier NM, Grizzle WE, Mikhail FM, Roth KA, Carroll SL. 2013. Transgenic mice overexpressing neuregulin-1 model neurofibroma-malignant peripheral nerve sheath tumor progression and implicate specific chromosomal copy number variations in tumorigenesis. <i>Am J Pathol.</i> Mar;182(3):646-67. doi: 10.1016/j.ajpath.2012.11.017. Epub 2013 Jan 13. PMID: 23321323; PMCID: PMC3586689. 2. Brosius SN, Turk AN, Byer SJ, Brossier NM, Kohli L, Whitmire A, Mikhail FM, Roth KA, Carroll SL. 2014. Neuregulin-1 overexpression and Trp53 haploinsufficiency cooperatively promote de novo malignant peripheral nerve sheath tumor pathogenesis. <i>Acta Neuropathol.</i> Apr;127(4):573-91. doi: 10.1007/s00401-013-1209-3. PMID: 24232507; PMCID: PMC3999224. 3. Brossier NM, Precht AM, Longo JF, Barnes S, Wilson LS, Byer SJ, Brosius SN, Carroll SL. 2015. Classic Ras proteins promote proliferation and survival via distinct phosphoproteome alterations in neurofibromin-null malignant peripheral nerve sheath tumor cells. <i>J Neuropathol Exp Neurol.</i> Jun;74(6):568-86. doi: 10.1097/NEN.0000000000000201. PMID: 25946318; PMCID: PMC4433624.*
NF130101	David Kirsch \$622,303.00	<ul style="list-style-type: none"> • Several cell lines developed from MPNSTs from in mice with NF1 haploinsufficient and NF1 wild-type stroma 	<ol style="list-style-type: none"> 1. Dodd RD, Lee CL, Overton T, Huang W, Eward WC, Luo L, Ma Y, Ingram DR, Torres KE, Cardona DM, Lazar AJ, Kirsch DG. 2017. NF1+/- hematopoietic cells accelerate malignant peripheral nerve sheath tumor development without altering chemotherapy response. <i>Cancer Res.</i> Aug 15;77(16):4486-4497. doi: 10.1158/0008-5472.CAN-16-2643. Epub 2017 Jun 23. PMID: 28646022; PMCID: PMC5839126. 2. Huang J, Chen M, Whitley MJ, Kuo HC, Xu ES, Walens A, Mowery YM, Van Mater D, Eward WC, Cardona DM, et al. 2017. Generation and comparison of CRISPR-Cas9 and Cre-mediated genetically engineered mouse models of sarcoma. <i>Nat Commun.</i> Jul 10;8:15999. doi: 10.1038/ncomms15999. PMID: 28691711; PMCID: PMC5508130.
NF140044	Cristina Fernandez-Valle \$777,059.00	<ul style="list-style-type: none"> • Human Schwann cell lines with NF2 shRNA mediated knock-down of merlin and lentiviral transduction of luciferase 	<ol style="list-style-type: none"> 1. Angus SP, Oblinger JL, Stuhlmiller TJ, DeSouza PA, Beauchamp RL, Witt L, Chen X, Jordan JT, Gilbert TSK, Stemmer-Rachamimov A, et al. 2018. EPH receptor signaling as a novel therapeutic target in NF2-deficient meningioma. <i>Neuro Oncol.</i> Aug 2;20(9):1185-1196. doi: 10.1093/neuonc/noy046. PMID: 29982664; PMCID: PMC6071664. 2. Synodos for NF2 Consortium, Allaway R, Angus SP, Beauchamp RL, Blakeley JO, Bott M, Burns SS, Carlstedt A, Chang LS, Chen X, et al. 2018. Traditional and systems biology based drug discovery for the rare tumor syndrome neurofibromatosis type 2. <i>PLoS One.</i> Jun 13;13(6):e0197350. doi: 10.1371/journal.pone.0197350. PMID: 29897904; PMCID: PMC5999111. 3. Fuse MA, Dinh CT, Vitte J, Kirkpatrick J, Mindos T, Plati SK, Young JI, Huang J, Carlstedt A, Franco MC, et al. 2019. Preclinical assessment of MEK1/2 inhibitors for neurofibromatosis type 2-associated schwannomas reveals differences in efficacy and drug resistance development. <i>Neuro Oncol.</i> Mar 18;21(4):486-497. doi: 10.1093/neuonc/noz002. PubMed PMID: 30615146; PubMed Central PMCID: PMC6422635.*

*Additional references can be found in Appendix A.

Cell Lines

CELL LINES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF140062	Ping Chi \$702,989.04	<ul style="list-style-type: none"> Isogenic MPNST cell lines that harbor NF1/CDKN2A loss or NF1/CDKN2A/PRC2 loss 	<ol style="list-style-type: none"> Prieto-Granada CN, Wiesner T, Messina JL, Jungbluth AA, Chi P, Antonescu CR. 2016. Loss of H3K27me3 expression is a highly sensitive marker for sporadic and radiation-induced MPNST. <i>Am J Surg Pathol.</i> Apr;40(4):479-89. doi: 10.1097/PAS.0000000000000564. PMID: 26645727; PMCID: PMC4882106. Moore AR, Ceraudo E, Sher JJ, Guan Y, Shoushtari AN, Chang MT, Zhang JQ, Walczak EG, Kazmi MA, Taylor BS, et al. 2016. Recurrent activating mutations of G-protein-coupled receptor CYSLTR2 in uveal melanoma. <i>Nat Genet.</i> Jun;48(6):675-80. doi: 10.1038/ng.3549. Epub 2016 Apr 18. PMID: 27089179; PMCID: PMC5032652. Lu C, Jain SU, Hoelper D, Bechet D, Molden RC, Ran L, Murphy D, Venneti S, Hameed M, Pawel BR, et al. 2016. Histone H3K36 mutations promote sarcomagenesis through altered histone methylation landscape. <i>Science.</i> May 13;352(6287):844-9. doi: 10.1126/science.aac7272. PMID: 27174990; PMCID: PMC4928577.*
NF150076	John Albeck \$706,437.00	<ul style="list-style-type: none"> Reporter cell line C8-D1A-EA, an immortalized astrocytic cell line expressing reporters for both ERK and Akt activity Reporter cell line C8-D1A-EA with shRNA targeted knockdown of NF1 	<ol style="list-style-type: none"> Gillies TE, Pargett M, Minguet M, Davies AE, Albeck JG. 2017. Linear integration of ERK activity predominates over persistence detection in Fra-1 regulation. <i>Cell Syst.</i> Dec 27;5(6):549-563.e5. doi: 10.1016/j.cels.2017.10.019. Epub 2017 Nov 29. PMID: 29199017; PMCID: PMC5746471. Pargett M, Albeck JG. 2018. Live-cell imaging and analysis with multiple genetically encoded reporters. <i>Curr Protoc Cell Biol.</i> Mar;78(1):4.36.1-4.36.19. doi: 10.1002/cpcb.38. PMID: 30040182; PMCID: PMC6060629. Gillies TE, Pargett M, Silva JM, Teragawa CK, McCormick F, Albeck JG. 2020. Oncogenic mutant RAS signaling activity is rescaled by the ERK/MAPK pathway. <i>Mol Syst Biol.</i> Oct;16(10):e9518. doi: 10.15252/msb.20209518. PMID: 33073539; PMCID: PMC7569415.*
NF150085	Hongjun Sun \$321,747.88	<ul style="list-style-type: none"> Validated iPSC lines from six NF1 patients of NF1 	
NF960013	George Devries \$481,542.00	<ul style="list-style-type: none"> Immortalized human NF1 MPNST cell line T265-2C 	<ol style="list-style-type: none"> Lee PR, Cohen JE, Tendi EA, Farrer R, DE Vries GH, Becker KG, Fields RD. 2004. Transcriptional profiling in an MPNST-derived cell line and normal human Schwann cells. <i>Neuron Glia Biol.</i> May;1(2):135-47. doi: 10.1017/s1740925x04000274. PMID: 16429615; PMCID: PMC1325299.

*Additional references can be found in Appendix A.

CELL LINES (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF980014	Ali Andalibi \$615,324.00	<ul style="list-style-type: none"> Immortalized human NF2 schwannoma line HEI-193 	<ol style="list-style-type: none"> Fraenzer JT, Pan H, Minimo L Jr, Smith GM, Knauer D, Hung G. 2003. Overexpression of the NF2 gene inhibits schwannoma cell proliferation through promoting PDGFR degradation. <i>Int J Oncol.</i> Dec;23(6):1493-500. PMID: 14612918. Hung G, Colton J, Fisher L, Oppenheimer M, Faudoa R, Slattery W, Linthicum F. 2002. Immunohistochemistry study of human vestibular nerve schwannoma differentiation. <i>Glia.</i> Jun;38(4):363-70. doi: 10.1002/glia.10077. PMID: 12007148. Hung G, Li X, Faudoa R, Xeu Z, Kluwe L, Rhim JS, Slattery W, Lim D. 2002. Establishment and characterization of a schwannoma cell line from a patient with neurofibromatosis 2. <i>Int J Oncol.</i> Mar;20(3):475-82. PMID: 11836557.*

*Additional references can be found in Appendix A.

Databases and Datasets

They are organized sets of data structured in a manner that can be utilized in different ways. NF-related data aids in comparison, prediction, quantitative/qualitative inference and analysis, adds to the breadth of information that promotes the research field.

DATABASES AND DATASETS

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF050145</u>	Victor-Felix Mautner \$628,550.00	<ul style="list-style-type: none"> Cohort of malignant peripheral nerve sheath tumors (MPNST) and plexiform neurofibromas (pNF) tumor samples 	<ol style="list-style-type: none"> Holtkamp N, Malzer E, Zietsch J, Okuducu AF, Mucha J, Mawrin C, Mautner VF, Schildhaus HU, von Deimling A. 2008. EGFR and erbB2 in malignant peripheral nerve sheath tumors and implications for targeted therapy. <i>Neuro Oncol.</i> Dec;10(6):946-57. doi: 10.1215/15228517-2008-053. Epub 2008 Jul 23. PMID: 18650488; PMCID: PMC2719009.
<u>NF050202</u>	Scott Plotkin \$1,454,775.00	<ul style="list-style-type: none"> Serum repository of the subjects enrolled in the protocol 	<ol style="list-style-type: none"> Smith MJ, Esparza S, Merker VL, Muzikansky A, Bredella MA, Harris GJ, Kassarian A, Cai W, Walker JA, Mautner VF, Plotkin SR. 2013. Plasma S100β is not a useful biomarker for tumor burden in neurofibromatosis. <i>Clin Biochem.</i> May;46(7-8):698-700. doi: 10.1016/j.clinbiochem.2012.12.007. Epub 2012 Dec 19. PMID: 23261835. Jaremko JL, MacMahon PJ, Torriani M, Merker VL, Mautner VF, Plotkin SR, Bredella MA. 2012. Whole-body MRI in neurofibromatosis: incidental findings and prevalence of scoliosis. <i>Skeletal Radiol.</i> Aug;41(8):917-23. doi: 10.1007/s00256-011-1333-x. Epub 2011 Dec 7. PMID: 22146869. Plotkin SR, Bredella MA, Cai W, Kassarian A, Harris GJ, Esparza S, Merker VL, Munn LL, Muzikansky A, Askenazi M, Nguyen R, Wenzel R, Mautner VF. 2012. Quantitative assessment of whole-body tumor burden in adult patients with neurofibromatosis. <i>PLoS One.</i> 7(4):e35711. doi: 10.1371/journal.pone.0035711. Epub 2012 Apr 27. PMID: 22558206; PMCID: PMC3338705.
<u>NF110080</u>	Carrie Bearden \$1,015,482.00	<ul style="list-style-type: none"> Biospecimens (DNA and PBMC samples) from study participants 	<ol style="list-style-type: none"> Tomson SN, Schreiner MJ, Narayan M, Rosser T, Enrique N, Silva AJ, Allen GI, Bookheimer SY, Bearden CE. 2015. Resting state functional MRI reveals abnormal network connectivity in neurofibromatosis 1. <i>Hum Brain Mapp.</i> Nov;36(11):4566-81. doi: 10.1002/hbm.22937. Epub 2015 Aug 25. PMID: 26304096; PMCID: PMC4619152. Jonas RK, Roh E, Montojo CA, et al. 2017. Risky decision making in Neurofibromatosis Type 1: An exploratory study. <i>Biol Psychiatry Cogn Neurosci Neuroimaging</i> 2(2):170-179. doi: 10.1016/j.bpsc.2016.12.003. PMID: 28736755 PMCID: PMC5518467.
<u>NF960039</u>	Mia MacCollin \$540,478.00	<ul style="list-style-type: none"> NF2 germline mutation map 	<ol style="list-style-type: none"> Egan RA, Thompson CR, MacCollin M, Lessell S. 2001. Monocular elevator paresis in neurofibromatosis type 2. <i>Neurology.</i> May 8;56(9):1222-4. doi: 10.1212/wnl.56.9.1222. PMID: 11342693. Jacoby LB, MacCollin M, Parry DM, Kluwe L, Lynch J, Jones D, Gusella JF. 1999. Allelic expression of the NF2 gene in neurofibromatosis 2 and schwannomatosis. <i>Neurogenetics.</i> Apr;2(2):101-8. doi: 10.1007/s100480050060. PMID: 10369886. Kluwe L, Mautner V, Parry DM, Jacoby LB, Baser M, Gusella J, Davis K, Stavrou D, MacCollin M. 2000. The parental origin of new mutations in neurofibromatosis 2. <i>Neurogenetics.</i> Sep;3(1):17-24. doi: 10.1007/s100480000088. PMID: 11085592.*

*Additional references can be found in Appendix A.

DATABASES AND DATASETS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF970002</u>	Bruce Korf \$997,500.00	<ul style="list-style-type: none"> Study data added to the NF International Database 	
<u>NF990008</u>	Andreas Kurtz \$309,760.00	<ul style="list-style-type: none"> A DNA repository has been established for NF1 subpopulations with high and low numbers of neurofibromas. 	<ol style="list-style-type: none"> Kurtz A, Lueth M, Kluwe L, Zhang T, Foster R, Mautner VF, Hartmann M, Tan DJ, Martuza RL, Friedrich RE, Driever PH, Wong LJ. 2004. Somatic mitochondrial DNA mutations in neurofibromatosis type 1-associated tumors. <i>Mol Cancer Res</i>. Aug;2(8):433-41. PMID: 15328370.
<u>NF990015</u>	Andre Bernards \$341,849.00	<ul style="list-style-type: none"> Genome Stability Gene SNP database listing information on 793 missense SNPs in 293 candidate genome stability genes 	
<u>NF000025</u>	Zygmunt Derewenda \$815,791.00	<ul style="list-style-type: none"> Deposited the crystal structure of the FERM domain of merlin into a protein data bank (PDB code 1H4R). Crystal structure of the PDZ tandem of human syntenin. Crystal structure analysis of the second PDZ domain of syntenin. Crystal structure of the complex of PDZ2 of syntenin with an interleukin 5 receptor peptide. Crystal structure of the complex of PDZ2 of syntenin with a syndecan-4 peptide. Crystal structure of the complex of the PDZ tandem of syntenin with an interleukin 5 receptor peptide. Ultra-high resolution crystal structure of syntenin PDZ2. Crystal structure of the PDZ tandem of human syntenin with TNEYKV peptide. The following structures will be deposited in the protein data bank: (1) PDZ tandem of syntenin with ESYF peptide, and (2) PDZ tandem of syntenin with TNEAYF peptide. 	<ol style="list-style-type: none"> Kang BS, Cooper DR, Jelen F, Devedjiev Y, Derewenda U, Dauter Z, Otlewski J, Derewenda ZS. 2003. PDZ tandem of human syntenin: crystal structure and functional properties. <i>Structure</i>. Apr;11(4):459-68. doi: 10.1016/s0969-2126(03)00052-2. PMID: 12679023. Kang BS, Cooper DR, Devedjiev Y, Derewenda U, Derewenda ZS. 2003. Molecular roots of degenerate specificity in syntenin's PDZ2 domain: reassessment of the PDZ recognition paradigm. <i>Structure</i>. Jul;11(7):845-53. doi: 10.1016/s0969-2126(03)00125-4. PMID: 12842047. Kang BS, Cooper DR, Devedjiev Y, Derewenda U, Derewenda ZS. 2002. The structure of the FERM domain of merlin, the neurofibromatosis type 2 gene product. <i>Acta Crystallogr D Biol Crystallogr</i>. Mar;58(Pt 3):381-91. doi: 10.1107/s0907444901021175. Epub 2002 Feb 21. PMID: 11856822.*
<u>NF010138</u>	Eric Hoffman \$449,919.00	<ul style="list-style-type: none"> Microarray data set from the hippocampus of mice with Nf1 haploinsufficiency 	<ol style="list-style-type: none"> Donarum EA, Halperin RF, Stephan DA, Narayanan V. 2006. Cognitive dysfunction in NF1 knock-out mice may result from altered vesicular trafficking of APP/DRD3 complex. <i>BMC Neurosci</i>. Mar 8;7:22. doi: 10.1186/1471-2202-7-22. PMID: 16524466; PMCID: PMC1434756.

*Additional references can be found in Appendix A.

Databases and Datasets

DATABASES AND DATASETS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF020035</u>	Andre Bernards \$1,416,876.00	<ul style="list-style-type: none"> Beagle Database: a genome-wide, multi-species biological annotation tool (incorporates published Ras superfamily database) 	<ol style="list-style-type: none"> Bernards A, Settleman J. 2004. GAP control: regulating the regulators of small GTPases. <i>Trends Cell Biol.</i> Jul;14(7):377-85. doi: 10.1016/j.tcb.2004.05.003. PMID: 15246431. Bernards A, Settleman J. 2005. GAPs in growth factor signalling. <i>Growth Factors.</i> Jun;23(2):143-9. doi: 10.1080/08977190500130480. PMID: 16019436. Bernards A. 2006. Ras superfamily and interacting proteins database. <i>Methods Enzymol.</i> 407:1-9. doi: 10.1016/S0076-6879(05)07001-1. PMID: 16757309. Bernards A, Settleman J. 2007. GEFs in growth factor signaling. <i>Growth Factors.</i> Oct;25(5):355-61. doi: 10.1080/08977190701830375. PMID: 18236214.
<u>NF043037</u>	Jeffrey Peterson \$500,000.00	<ul style="list-style-type: none"> Electronic database in ChemFinder (Cambridgesoft) containing structures and biophysical data on 30,000 small molecules and their activity in the Pak1 inhibitor screen. 	<ol style="list-style-type: none"> Eswaran J, Lee WH, Debreczeni JE, Filippakopoulos P, Turnbull A, Fedorov O, Deacon SW, Peterson JR, Knapp S. 2007. Crystal Structures of the p21-activated kinases PAK4, PAK5, and PAK6 reveal catalytic domain plasticity of active group II PAKs. <i>Structure.</i> Feb;15(2):201-13. doi: 10.1016/j.str.2007.01.001. PMID: 17292838; PMCID: PMC1885963. Rennefahrt UE, Deacon SW, Parker SA, Devarajan K, Beeser A, Chernoff J, Knapp S, Turk BE, Peterson JR. 2007. Specificity profiling of Pak kinases allows identification of novel phosphorylation sites. <i>J Biol Chem.</i> May 25;282(21):15667-78. doi: 10.1074/jbc.M700253200. Epub 2007 Mar 28. PMID: 17392278. Deacon SW, Peterson JR. 2008. Chemical inhibition through conformational stabilization of Rho GTPase effectors. <i>Handb Exp Pharmacol.</i> 186:431-60. doi: 10.1007/978-3-540-72843-6_18. PMID: 18491063.*
<u>NF050018</u>	John Mulvihill \$761,215.00	<ul style="list-style-type: none"> Cohort of US mortality data on individuals diagnosed with NF1. Repository of blood samples from consented NF1 participants. 	
<u>NF050110</u>	Arie Perry \$99,700.20	<ul style="list-style-type: none"> SNP array from MPNSTs 	<ol style="list-style-type: none"> Dorsi MJ, Chen L, Murinson BB, Pogatzki-Zahn EM, Meyer RA, Belzberg AJ. 2008. The tibial neuroma transposition (TNT) model of neuroma pain and hyperalgesia. <i>Pain.</i> Feb;134(3):320-334. doi: 10.1016/j.pain.2007.06.030. Epub 2007 Aug 27. PMID: 17720318.

*Additional references can be found in Appendix A.

DATABASES AND DATASETS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF093077</u>	Subbaya Subramanian \$601,550.00	<ul style="list-style-type: none"> • microRNA profile of neurofibromas, schwannomas, and peripheral nerve sheath tumors from humans and mice 	<ol style="list-style-type: none"> 1. Sarver AL, Subramanian S. 2012. Competing endogenous RNA database. <i>Bioinformatics</i>. 8(15):731-3. doi: 10.6026/97320630008731. Epub Aug 3. PMID: 23055620; PMCID: PMC3449376. 2. Subramanian S, Kartha RV. 2012. MicroRNA-mediated gene regulations in human sarcomas. <i>Cell Mol Life Sci</i>. Nov;69(21):3571-85. doi: 10.1007/s00018-012-1127-x. Epub Aug 25. PMID: 22922987. 3. Shu J, Xia Z, Li L, Liang ET, Slipek N, Shen D, Foo J, Subramanian S, Steer CJ. 2012. Dose-dependent differential mRNA target selection and regulation by let-7a-7f and miR-17-92 cluster microRNAs. <i>RNA Biol</i>. Oct;9(10):1275-87. doi: 10.4161/ma.21998. Epub 2012 Sep 20. PMID: 22995834; PMCID: PMC3583858.
<u>NF140039</u>	Tina Izard \$1,086,597.00	<ul style="list-style-type: none"> • Crystal structures of NF2 mutant proteins 	<ol style="list-style-type: none"> 1. Chinthalapudi K, Mandati V, Zheng J, Sharff AJ, Bricogne G, Griffin PR, Kissil J, Izard T. 2018. Lipid binding promotes the open conformation and tumor-suppressive activity of neurofibromin 2. <i>Nat Commun</i>. Apr 6;9(1):1338. doi: 10.1038/s41467-018-03648-4. PMID: 29626191; PMCID: PMC5889391.

Methods

Methods are standardized processes of carrying out tasks required to achieve certain goals. NF research methods improve the scientific and systematic courses of action that scientists follow in their tasks.

METHODS

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF020064</u>	David Muir \$1,548,903.00	<ul style="list-style-type: none"> Developed an in vivo delivery system for delivering AAV-endostatin to NF1 tumor xenografts following subcutaneous injection of a 293/AAV-endostatin cell line. 	<ol style="list-style-type: none"> Wu M, Wallace MR, Muir D. 2005. Tumorigenic properties of neurofibromin-deficient Schwann cells in culture and as syngrafts in Nf1 knockout mice. <i>J Neurosci Res</i>. Nov 1;82(3):357-67. doi: 10.1002/jnr.20646. PMID: 16180234. Wu M, Wallace MR, Muir D. 2006. Nf1 haploinsufficiency augments angiogenesis. <i>Oncogene</i>. Apr 13;25(16):2297-303. doi: 10.1038/sj.onc.1209264. PMID: 16288202. Fishbein L, Zhang X, Fisher LB, Li H, Campbell-Thompson M, Yachnis A, Rubenstein A, Muir D, Wallace MR. 2007. In vitro studies of steroid hormones in neurofibromatosis 1 tumors and Schwann cells. <i>Mol Carcinog</i>. Jul;46(7):512-23. doi: 10.1002/mc.20236. PMID: 17393410.*
<u>NF020086</u>	Westley Friesen \$614,740.00	<ul style="list-style-type: none"> Developed assay for detection of neurofibromin by immunofluorescence. Synthesis of PTC-1a, a proprietary compound. 	
<u>NF030032</u>	James Gusella \$150,000.00	<ul style="list-style-type: none"> Developed methodology for the collection, digitalization, and volumetric analysis of magnetic resonance images. 	
<u>NF050028</u>	David Gutmann \$1,956,438.00	<ul style="list-style-type: none"> Non-invasive technique to detect optic glioma in mouse model of NF1 	<ol style="list-style-type: none"> Banerjee D, Hegedus B, Gutmann DH, Garbow JR. 2007. Detection and measurement of neurofibromatosis-1 mouse optic glioma in vivo. <i>Neuroimage</i>. May 1;35(4):1434-7. doi: 10.1016/j.neuroimage.2007.02.019. Epub 2007 Feb 23. PMID: 17383899; PMCID: PMC2735870. Hegedus B, Banerjee D, Yeh TH, Rothermich S, Perry A, Rubin JB, Garbow JR, Gutmann DH. 2008. Preclinical cancer therapy in a mouse model of neurofibromatosis-1 optic glioma. <i>Cancer Res</i>. Mar 1;68(5):1520-8. doi: 10.1158/0008-5472.CAN-07-5916. PMID: 18316617. Banerjee S, Byrd JN, Gianino SM, Harpstrite SE, Rodriguez FJ, Tuskan RG, Reilly KM, Piwnica-Worms DR, Gutmann DH. 2010. The neurofibromatosis type 1 tumor suppressor controls cell growth by regulating signal transducer and activator of transcription-3 activity in vitro and in vivo. <i>Cancer Res</i>. Feb 15;70(4):1356-66. doi: 10.1158/0008-5472.CAN-09-2178. Epub 2010 Feb 2. PMID: 20124472; PMCID: PMC5534849.*
<u>NF050050</u>	Kevin McHugh \$759,338.25	<ul style="list-style-type: none"> Immunohistochemical detection of neurofibromin in normal mouse bone and in a mouse model of fracture healing. 	

*Additional references can be found in Appendix A.

METHODS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF050137</u>	Norbert Perrimon \$99,663.08	<ul style="list-style-type: none"> Technique to quantify phenotypic cell profiles facilitating screening for modifiers of NF1 and NF2 phenotypes 	<ol style="list-style-type: none"> Bakal C, Aach J, Church G, Perrimon N. 2007. Quantitative morphological signatures define local signaling networks regulating cell morphology. <i>Science</i>. Jun 22;316(5832):1753-6. doi: 10.1126/science.1140324. PMID: 17588932. Nir O, Bakal C, Perrimon N, Berger B. 2010. Inference of RhoGAP/ GTPase regulation using single-cell morphological data from a combinatorial RNAi screen. <i>Genome Res</i>. Mar;20(3):372-80. doi: 10.1101/gr.100248.109. Epub 2010 Feb 9. PMID: 20144944; PMCID: PMC2840989.
<u>NF050184</u>	Andrew Hoffman \$1,425,614.00	<ul style="list-style-type: none"> Associated chromosome trap (ACT) assay 	<ol style="list-style-type: none"> Ling JQ, Hoffman AR. 2007. Epigenetics of long-range chromatin interactions. <i>Pediatr Res</i>. May;61(5 Pt 2):11R-16R. doi: 10.1203/pdr.Ob013e31804575db. PMID: 17413850. Ling JQ, Hou A, Hoffman AR. 2011. Long-range DNA interactions are specifically altered by locked nucleic acid-targeting of a CTCF binding site. <i>Biochim Biophys Acta</i>. Jan;1809(1):24-33. doi: 10.1016/j.bbagr.2010.11.004. Epub 2010 Nov 24. PMID: 21111075. Ling J, Hoffman AR. 2011. Associated chromosome trap for identifying long-range DNA interactions. <i>J Vis Exp</i>. Apr 23;(50):2621. doi: 10.3791/2621. PMID: 21540824; PMCID: PMC3169263.
<u>NF080089</u>	Steven Carroll \$757,623.48	<ul style="list-style-type: none"> Orthotopic xenografting method for luciferase-tagged MPNST cells 	<ol style="list-style-type: none"> Byer SJ, Eckert JM, Brossier NM, Clodfelder-Miller BJ, Turk AN, Carroll AJ, Kappes JC, Zinn KR, Prasain JK, Carroll SL. 2011. Tamoxifen inhibits malignant peripheral nerve sheath tumor growth in an estrogen receptor-independent manner. <i>Neuro Oncol</i>. Jan;13(1):28-41. doi: 10.1093/neuonc/noq146. Epub 2010 Nov 12. PMID: 21075781; PMCID: PMC3018903. Brossier NM, Byer SJ, Peavler LT, Carroll SL. 2012. ErbB membrane tyrosine kinase receptors: Analyzing migration in a highly complex signaling system. <i>Protein Kinase Technologies</i>. 02 May; 68: 105-131. https://doi.org/10.1007/978-1-61779-824-5_7. Turk AN, Byer SJ, Zinn KR, Carroll SL. 2011. Orthotopic xenografting of human luciferase-tagged malignant peripheral nerve sheath tumor cells for in vivo testing of candidate therapeutic agents. <i>J Vis Exp</i>. Mar 7;(49):2558. doi: 10.3791/2558. PMID: 21460792; PMCID: PMC3197311.*
<u>NF093033</u>	David Gutmann \$855,850.41	<ul style="list-style-type: none"> PET technique to detect and evaluate promising agents for attention deficit in Nf1 mutant mice 	<ol style="list-style-type: none"> Brown JA, Xu J, Diggs-Andrews KA, Wozniak DF, Mach RH, Gutmann DH. 2011. PET imaging for attention deficit preclinical drug testing in neurofibromatosis-1 mice. <i>Exp Neurol</i>. Dec;232(2):333-8. doi: 10.1016/j.expneurol.2011.09.005. Epub 2011 Sep 22. PMID: 21963652; PMCID: PMC3202049. Gutmann DH, Parada LF, Silva AJ, Ratner N. 2012. Neurofibromatosis type 1: modeling CNS dysfunction. <i>J Neurosci</i>. Oct 10;32(41):14087-93. doi: 10.1523/JNEUROSCI.3242-12.2012. PMID: 23055477; PMCID: PMC3477849. Diggs-Andrews KA, Gutmann DH. 2013. Modeling cognitive dysfunction in neurofibromatosis-1. <i>Trends Neurosci</i>. Apr;36(4):237-47. doi: 10.1016/j.tins.2012.12.002. Epub 2013 Jan 8. PMID: 23312374; PMCID: PMC3622809.*

*Additional references can be found in Appendix A.

Methods

METHODS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF093047</u>	Fernando Camargo \$690,666.00	<ul style="list-style-type: none"> • TEAD-based cell lines and screening technique 	<ol style="list-style-type: none"> 1. Ramos A, Camargo FD. 2012. The Hippo signaling pathway and stem cell biology. <i>Trends Cell Biol.</i> Jul;22(7):339-46. doi: 10.1016/j.tcb.2012.04.006. Epub 2012 May 31. PMID: 22658639; PMCID: PMC3383919. 2. Tremblay AM, Camargo FD. 2012. Hippo signaling in mammalian stem cells. <i>Semin Cell Dev Biol.</i> Sep;23(7):818-26. doi: 10.1016/j.semcd.2012.08.001. Epub 2012 Aug 8. PMID: 23034192. 3. Mohseni M, Sun J, Lau A, Curtis S, Goldsmith J, Fox VL, Wei C, Frazier M, Samson O, Wong KK, Kim C, Camargo FD. 2014. A genetic screen identifies an LKB1-MARK signalling axis controlling the Hippo-YAP pathway. <i>Nat Cell Biol.</i> Jan;16(1):108-17. doi: 10.1038/ncb2884. Epub 2013 Dec 22. Erratum in: <i>Nat Cell Biol.</i> 2014 Feb;16(2):200. Wong, Kwok-Kim [corrected to Wong, Kwok-Kin]. PMID: 24362629; PMCID: PMC4159053.*
<u>NF100148</u>	Maura Cosetti \$107,956.00	<ul style="list-style-type: none"> • QoL instrument for NF patients 	<ol style="list-style-type: none"> 1. Cosetti MK, Golfinos JG, Roland JT Jr. 2015. Quality of Life (QoL) Assessment in Patients with Neurofibromatosis Type 2 (NF2). <i>Otolaryngol Head Neck Surg.</i> Oct;153(4):599-605. doi: 10.1177/0194599815573002. Epub 2015 Mar 16. PMID: 25779467.
<u>NF130061</u>	Cathie Pflieger \$675,318.00	<ul style="list-style-type: none"> • Method to examine the lymph gland and hemocytes in <i>Drosophila</i> larvae 	<ol style="list-style-type: none"> 1. Wolman MA, de Groh ED, McBride SM, Jongens TA, Granato M, Epstein JA. 2014. Modulation of cAMP and ras signaling pathways improves distinct behavioral deficits in a zebrafish model of neurofibromatosis type 1. <i>Cell Rep.</i> Sep 11;8(5):1265-70. doi: 10.1016/j.celrep.2014.07.054. Epub 2014 Aug 28. PMID: 25176649; PMCID: PMC5850931. 2. Fagan JK, Dollar G, Lu Q, Barnett A, Pechuan Jorge J, Schlosser A, Pflieger C, Adler P, Jenny A. 2014. Combover/CG10732, a novel PCP effector for <i>Drosophila</i> wing hair formation. <i>PLoS One.</i> Sep 10;9(9):e107311. doi: 10.1371/journal.pone.0107311. PMID: 25207969; PMCID: PMC4160248. 3. Reimels TA, Pflieger CM. 2015. <i>Drosophila</i> Rabex-5 restricts Notch activity in hematopoietic cells and maintains hematopoietic homeostasis. <i>J Cell Sci.</i> Dec 15;128(24):4512-25. doi: 10.1242/jcs.174433. Epub 2015 Nov 13. PMID: 26567216; PMCID: PMC4696494.*
<u>NF150076</u>	John Albeck \$706,437.00	<ul style="list-style-type: none"> • Gene delivery method to generate populations of primary fibroblasts expressing a FRET-based ERK reporter • Method for performing quantitative live-cell analysis of kinetic trajectories from individual cells 	<ol style="list-style-type: none"> 1. Gillies TE, Pargett M, Minguet M, Davies AE, Albeck JG. 2017. Linear Integration of ERK Activity Predominates over Persistence Detection in Fra-1 Regulation. <i>Cell Syst.</i> Dec 27;5(6):549-563. e5. doi: 10.1016/j.cels.2017.10.019. Epub 2017 Nov 29. PMID: 29199017; PMCID: PMC5746471. 2. Pargett M, Albeck JG. 2018. Live-Cell Imaging and Analysis with Multiple Genetically Encoded Reporters. <i>Curr Protoc Cell Biol.</i> Mar;78(1):4.36.1-4.36.19. doi: 10.1002/cpcb.38. PMID: 30040182; PMCID: PMC6060629. 3. Gillies TE, Pargett M, Silva JM, Teragawa CK, McCormick F, Albeck JG. 2020. Oncogenic mutant RAS signaling activity is rescaled by the ERK/MAPK pathway. <i>Mol Syst Biol.</i> Oct;16(10):e9518. doi: 10.15252/msb.20209518. PMID: 33073539; PMCID: PMC7569415.

*Additional references can be found in Appendix A.

METHODS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF960027</u>	Margaret Wallace \$594,663.00	Method to culture and xenograft human neurofibromin-deficient Schwann cells: <ul style="list-style-type: none"> Polyclonal rabbit anti-neurofibromin: PcNF27 Monoclonal rabbit anti-neurofibromin: McNF27a and McNF27b 	<ol style="list-style-type: none"> Muir D, Neubauer D, Lim IT, Yachnis AT, Wallace MR. 2001. Tumorigenic properties of neurofibromin-deficient neurofibroma Schwann cells. <i>Am J Pathol.</i> Feb;158(2):501-13. doi: 10.1016/S0002-9440(10)63992-2. PMID: 11159187; PMCID: PMC1850316.
<u>NF960048</u>	Kevin Shannon \$946,888.00	<ul style="list-style-type: none"> The studies conducted to date have established a regimen for administering cyclophosphamide with and without radiation to F1 C56BL6/129Sv mice that should be useful for future studies of tumorigenesis and chemopreventive strategies in Nf1 mice. The Hpvt 	<ol style="list-style-type: none"> Weiss B, Bollag G, Shannon K. 1999. Hyperactive Ras as a therapeutic target in neurofibromatosis type 1. <i>Am J Med Genet.</i> Mar 26;89(1):14-22. PMID: 10469432.
<u>NF970002</u>	Bruce Korf \$997,500.00	<ul style="list-style-type: none"> Volumetric MRI for measuring growth of plexiform neurofibromas 	<ol style="list-style-type: none"> Mautner VF, Friedrich RE, von Deimling A, Hagel C, Korf B, Knöfel MT, Wenzel R, Fünsterer C. 2003. Malignant peripheral nerve sheath tumours in neurofibromatosis type 1: MRI supports the diagnosis of malignant plexiform neurofibroma. <i>Neuroradiology.</i> Sep;45(9):618-25. doi: 10.1007/s00234-003-0964-6. Epub 2003 Jul 24. PMID: 12898075. Friedrich RE, Korf B, Fünsterer C, Mautner VF. 2003. Growth type of plexiform neurofibromas in NF1 determined on magnetic resonance images. <i>Anticancer Res.</i> Mar-Apr;23(2A):949-52. PMID: 12820328. Kluwe L, Friedrich RE, Korf B, Fahsold R, Mautner VF. 2002. NF1 mutations in neurofibromatosis 1 patients with plexiform neurofibromas. <i>Hum Mutat.</i> Mar;19(3):309. doi: 10.1002/humu.9018. PMID: 11857752.*
<u>NF990013</u>	Xandra Breakfield \$1,009,421.00	<ul style="list-style-type: none"> Novel method to image apoptosis in vivo 	<ol style="list-style-type: none"> Messerli SM, Tang Y, Giovannini M, Bronson R, Weissleder R, Breakefield XO. 2002. Detection of spontaneous schwannomas by MRI in a transgenic murine model of neurofibromatosis type 2. <i>Neoplasia.</i> Nov-Dec;4(6):501-9. doi: 10.1038/sj.neo.7900265. PMID: 12407444; PMCID: PMC1503664. Tang Y, Shah K, Messerli SM, Snyder E, Breakefield X, Weissleder R. 2003. In vivo tracking of neural progenitor cell migration to glioblastomas. <i>Hum Gene Ther.</i> Sep 1;14(13):1247-54. doi: 10.1089/104303403767740786. PMID: 12952596. Prabhakar S, Messerli SM, Stemmer-Rachamimov AO, Liu TC, Rabkin S, Martuza R, Breakefield XO. 2007. Treatment of implantable NF2 schwannoma tumor models with oncolytic herpes simplex virus G47Delta. <i>Cancer Gene Ther.</i> May;14(5):460-7. doi: 10.1038/sj.cgt.7701037. Epub 2007 Feb 16. PMID: 17304235.*

*Additional references can be found in Appendix A.

Methods

METHODS (CONT.)

Award Number	Principal Investigator Award Amount	Products or Resources	References
<u>NF990031</u>	Karen Stephens \$762,231.00	<ul style="list-style-type: none"> Developed a competitive, quantitative PCR method for differentiating one versus two copies of NF1 gene intron 31. 	<ol style="list-style-type: none"> Stephens K. 2003. Genetics of neurofibromatosis 1-associated peripheral nerve sheath tumors. <i>Cancer Invest.</i> 21(6):897-914. doi: 10.1081/cnv-120025093. PMID: 14735694. López-Correa C, Dorschner M, Brems H, Lázaro C, Clementi M, Upadhyaya M, Dooijes D, Moog U, Kehrer-Sawatzki H, Rutkowski JL, et al. 2001. Recombination hotspot in NF1 microdeletion patients. <i>Hum Mol Genet.</i> Jun 15;10(13):1387-92. doi: 10.1093/hmg/10.13.1387. PMID: 11440991. Messiaen L, Riccardi V, Peltonen J, Maertens O, Callens T, Karvonen SL, Leisti EL, Koivunen J, Vandenbroucke I, Stephens K, Pöyhönen M. 2003. Independent NF1 mutations in two large families with spinal neurofibromatosis. <i>J Med Genet.</i> Feb;40(2):122-6. doi: 10.1136/jmg.40.2.122. PMID: 12566521; PMCID: PMC1735368.*
<u>NF990038</u>	Harry Joe \$199,001.00	<ul style="list-style-type: none"> Statistical models and software package for analysis of familial data of various types (binary, count, continuous, censored). Software written in C programming languages, developed in Unix/Linux, runs also in Windows with Cygnus/Gnuwin. 	<ol style="list-style-type: none"> Baser ME, Friedman JM, Aeschliman D, Joe H, Wallace AJ, Ramsden RT, Evans DG. 2002. Predictors of the risk of mortality in neurofibromatosis 2. <i>Am J Hum Genet.</i> Oct;71(4):715-23. doi: 10.1086/342716. Epub 2002 Aug 22. PMID: 12235555; PMCID: PMC378530. Baser ME, Friedman JM, Wallace AJ, Ramsden RT, Joe H, Evans DG. 2002. Evaluation of clinical diagnostic criteria for neurofibromatosis 2. <i>Neurology.</i> Dec 10;59(11):1759-65. doi: 10.1212/01.wnl.0000035638.74084.f4. PMID: 12473765. Baser ME, Kuramoto L, Joe H, Friedman JM, Wallace AJ, Ramsden RT, Evans DG. 2003. Genotype-phenotype correlations for cataracts in neurofibromatosis 2. <i>J Med Genet.</i> Oct;40(10):758-60. doi: 10.1136/jmg.40.10.758. PMID: 14569124; PMCID: PMC1735286.*

*Additional references can be found in Appendix A.

Yeast Strains

A type of fungus that reproduces by budding and is useful for fermenting sugars. Yeast cell pathways have similarities with mammalian cells and can be used in the study of NF-mutations to support the understanding of cellular function.

YEAST STRAINS

Award Number	Principal Investigator Award Amount	Products or Resources	References
NF073046	Aaron Gitler \$452,000.00	<ul style="list-style-type: none">S. cerevisiae IRA strains: 1. 1Δ mutant, 2. ira2Δ mutant	



Appendix A: Additional References

MOUSE MODELS

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Appendix A: Additional References

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