



NF Clinic Network (NFCN) Application Form*

Clinic Name:

Texas Children's Hospital Neurofibromatosis Clinic

Affiliated Hospital:

Texas Children's Hospital

Affiliated University or Institution:

Baylor College of Medicine

Clinic Address:

Texas Children's Hospital Clinical Care Center
6621 Fannin, CC 1560
Houston, TX 77030

Clinic Director:

Sharon Plon, MD, Ph.D.

Clinic Coordinator Name:

Katie S. Plunkett, MS

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



The Neurofibromatosis Clinic Network (NFCN)

FORM PART A: Affiliate Clinic Application

1. ABOUT YOUR NF CLINIC

a. Is your NF Clinic:

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

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

The Neurofibromatosis Clinic at Texas Children's Hospital is a multidisciplinary clinic staffed by board certified clinical geneticists and post-doctoral fellows in clinical genetics which has been in operation for over twenty years. The coordinator is a board-certified genetic counselor and we provide multidisciplinary services and coordination of care for families (both adults and children) with Neurofibromatosis 1, Neurofibromatosis 2, Tuberous sclerosis, and von Hippel Lindau disease. The clinic meets once monthly.

2. CLINIC DIRECTOR and STAFF EXPERTISE

a. CLINIC DIRECTOR: Please describe:

- i. Your experience to date with NF care: Dr. Plon, a board-certified medical geneticist, has been the Chief of the Baylor College of Medicine/Texas Children's Hospital Neurofibromatosis Clinic since 1995. This is a monthly clinic that provides multi-disciplinary care for patients with diagnoses of NF1 and NF2. Dr. Plon has been providing clinical care to the patients seen in the clinic as well as participating in a number of local medical conferences, e.g. neuro-oncology tumor board where management decisions about patient with NF are made.
- ii. Your past and current association with NF clinical trials: Dr. Plon participated in the Plexiform Neurofibroma Natural History study. Dr. Plon also refers appropriate patients to Dr. Susan Blaney at the Texas Children's Cancer Center who directs the Phase I clinical trials including participation in the farnesyl transferase inhibitor study. Dr. Plon and the Solid Tumor team at Texas Children's Cancer Center will also be participating in the Phase II trial of Neoadjuvant Chemotherapy in Sporadic and Neurofibromatosis Type 1 associated High Grade Unresectable Malignant Peripheral Nerve Sheath Tumors.
- iii. Your past and current association with other clinical trials e.g. oncology trials: Dr. Plon does not have direct participation in treatment trials. For this purpose the clinic collaborates with Drs. Murali Chintagumpala, Alberto Pappo and Susan Blaney of the Texas Children's Cancer Center who have extensive experience in conducting chemotherapy clinical trials. Dr. Plon has participated in a number of cancer genetics clinical studies which include elucidating the genetic basis of families with inherited susceptibility to adult and pediatric cancers. Dr. Plon has also participated in studies to determine the impact of implementing genetic testing for cancer susceptibility.

b. CLINIC DIRECTOR: Please provide information on:

- i. Present and past funding you have received for NF research. Include funding source, date received, amount and project description.
Dr. Plon has not received funding directly for NF research.
- ii. Your NF-related clinical and scientific publications. Include Journal, Citation and Title.

Airewele, G.E., Sigurdson, A.J., Wiley, K.J., Frieden, B.E., Caldarera, L.W., Riccardi, V.M., Lewis, R.A., Chintagumpala, M.M., Ater, J.L., **Plon, S.E.**, and M.L. Bondy (2001). Neoplasms in Neurofibromatosis Type 1 are Related to Gender and Inheritance but not Family History of Cancer. *Genetic Epidemiology*, **20**:75-86.

Blazo, M .A., Lewis, R.A., Chintagumpala, M.M., Frazier M., McCluggage, C. and **S.E. Plon** (2004). Outcomes of Systematic Screening for Optic Pathway Tumors in Children with Neurofibromatosis Type I. *American J Medical Genetics*, *127A*(3):224-9.

Plon, S.E., and V.M. Riccardi (1999). “The Phakomatoses and Other Neurocutaneous Syndromes” in Oski’s Pediatrics: Principles and Practice, 3rd Edition, (Eds. Frank A. Oski et al.) J. B. Lippincott Company, Philadelphia, 417: 2019 - 2025.

Plon, S.E. (2005). “The Phakomatoses and Other Neurocutaneous Syndromes” in Oski’s Pediatrics: Principles and Practice, 4th Edition, (Eds. Frank A. Oski et al.) J. B. Lippincott Company, Philadelphia.

Plon, S.E. and M.A. Blazo (2002). Neurofibromatosis I, in, *UpToDate in Pediatrics*. - updated annually

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

1) Ms. Katie S. Plunkett, M.S.; Genetic Counselor; Coordinator and advisor to scheduling of monthly clinics; liaison to subspecialist services; contact person for physician and patient referrals to the Neurofibromatosis clinic.

2) Richard A. Lewis, M.D.: Dr. Lewis is a world-recognized ophthalmologist with extensive (over twenty-five years) experience in the treatment of patients with genetic eye diseases. He has continuously attended in our NF Clinic since coming to Baylor in the 1970’s. He is a member of the Advisory Board of the National Eye Institute as well as serving on the advisory board of many genetic disease support groups including the VHL Family Alliance.

3) Maria Blazo, M.D.: Dr. Blazo had her original postgraduate training in Family Medicine. She subsequently completed the Medical Genetics fellowship at Baylor College of Medicine. During this fellowship, she focused on clinical research related to the question of appropriate screening of young patients with NF1. Since completing the fellowship, Dr. Blazo now attends regularly in our NF Clinic, our Adult Genetics Clinic (where many adults with NF are diagnosed) and has an active Family Medicine clinical practice. Dr. Blazo attending the CTF clinical meetings this June in order to keep our clinic abreast of any changes in treatment guidelines for NF patients.

4) John Oghalai, M.D.: Dr. Oghalai is a board-certified otolaryngologist who was recruited to the Baylor faculty five years ago after completing his training at the University of California San Francisco. He has become an active member of our NF Clinic staff and participates in the care of NF2 patients with acoustic neuromas and

other facial tumors as well as NF1 patients with plexiform neurofibromas of the head and neck. His clinical practice includes both pediatric and adult patients.

5) Ms. San Juana Galvan; Clinic secretary; Assistant coordinator in scheduling patients for Neurofibromatosis clinic; organizes patient summaries; contacts patients in preparation for upcoming visits to facilitate referrals/subspecialist appointments; implements physician orders for patient care; sends correspondence to referring physicians and ensures follow-up care appointments are communicated to patients, families and referring physician.

6) Ms. Stephanie Gutierrez, B.A.; database manager; research coordinator and data manager for registry of NF patients enrolled in ongoing clinical trials; facilitates informed consent forms for Neurofibromatosis trials and information on patient registry.

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

Katie S. Plunkett, M.S., Genetic Counselor, is current clinic coordinator for NF patient services. Her duties include ensuring patients are scheduled appropriately for clinic in a timely manner, communicating with patients and families on the coordination of care in the Neurofibromatosis clinic, and assisting in patient care directly with genetic counseling; She also provides updated information resources to patients and families.

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

The Neurofibromatosis clinic has broad based access to expertise in areas of neuro-developmental pediatrics, neurology, neurosurgery, ophthalmology, oncology and dermatology. Research interests have historically included cancer family history, optic glioma, and skeletal manifestations in patients with Neurofibromatosis type I. Dr. Richard Lewis is an ophthalmologist with over twenty-five years of experience evaluating patients with NF1 for ophthalmic manifestations and attends in our clinic. Dr. Brendan Lee is a board-certified medical geneticist who has carried out clinical studies of bone abnormalities in NF1 patients as a collaboration between the NF1 Clinic and the Skeletal Dysplasia Clinic.

3. PATIENT SCHEDULING and REFERRALS

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

Patients overall receive a response to request for scheduling within two (2) days. Patients are scheduled for the next available clinic appointment generally within a 3 month time frame. Patients who require more urgent follow-up are contacted by physician staff (geneticist) to arrange alternate scheduling of appointment in either adult or pediatric genetics clinic.

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

PEDIATRIC CARE

SPECIALTY	DOCTOR	CLINIC ADDRESS	PHONE	EMAIL (if available)
Genetics	Dr. Sharon Plon, MD, PhD Dr. Maria Blazo, MD Dr. Christine Eng, MD	Clinical Care Center (CCC) Texas Children's Hospital 6701 Fannin St. 16 th Floor Houston, Tx 77030	832.822.4280	splon@bcm.edu ceng@bcm.edu mblazo@bcm.edu
Neurology	Dr. Marvin Fishman, MD	CCC 6701 Fannin 9 th Floor Ste. 950 Houston, TX 77030	832.822.1750	mfishman@bcm.edu
Orthopedics	Dr. William Phillips, MD	CCC 6701, Fannin 8 th Floor Ste. 0820.01 Houston, TX 77030	832.822.3100	willamp@bcm.edu
Developmental pediatrics/learning disabilities	Meyer Learning Center Isabella R.lovino	CCC 6701 Fannin 16 th Floor Houston, TX 77030	832.822.3400	iriovino@texaschildrens hospital.org
Ophthalmology	Dr. Richard Lewis, MD	Smith Tower 15 th . Floor Ste. 1501 Houston, TX 77030	713.798.6100	rlewis@bcm.edu
Neurosurgery	Dr. Dauser, MD	CCC 6701 Fannin, 9 th Floor Houston, TX 77030	832.822.3950	rdauser@bcm.edu
Plastic surgery	Dr. Hollier, MD	CCC 6701 Fannin 17 Floor	832.822.3180	larryh@bcm.tmc.edu
Neurooncology	Dr. Murali Chintagumpala, MD	Texas Children's Cancer Center 6701 Fannin, 14 th Floor, Ste. cc1410 Houston, TX 77030	832-822-4242	muralic@txccc.org

Medical Oncology/Radiation Oncology	Dr. Susan Blaney, MD/ Dr. David Kornguth, MD	Texas Children's Cancer Center/ MD Anderson Cancer Center 1515 Holcombe Blvd, Houston, TX 77030	832-822-4242/ 1-800-392-1611	smblaney@txccc.org dkorngut@mdanderson.org
Endocrinology	Dr. Karaviti, MD	CCC 6701 Fannin 11 th Floor, Ste. CC1130.03 Houston, TX 77030	832.822.3670	LPKaravi@texaschildrenshospital.org
Audiology/ENT	Dr. John Oghalai, MD	CCC 6701 Fannin 15 th Floor, Ste. CC520.30 Houston, TX 77030	832.822.3250	jso@bcm.edu
Radiology/Neuroradiology	Dr. Jill Hunter, MD	West Tower 6621 Fannin Floor Houston, TX 77030	832.824.5324	Jhunter@bcm.edu
General Surgery/Surgical Oncology	Dr. Jed Nuchtern, MD	CCC 6701 Fannin 8 th Floor Houston, TX 77030	832.822.3463	nuchtern@bcm.edu
Dermatology	Dr. Moise Levy, MD	CCC 6701 Fannin 8 th Floor, Ste. CC830.15 Houston, TX 77030	832.822.3463	levy@bcm.edu
Cardiovascular Disease	Dr. Wayne Franklin, MD Dr. Jeff Towbin, MD	West Tower 6621 Fannin 20th Floor, WT19-345H Houston, TX 77030	832.826.5731	wjf@bcm.edu jtowbin@bcm.edu
Oral and Maxillofacial Surgery	Dr. John Oghalai, MD Dr. Samuel Stal, MD	CCC 6701 Fannin 17 th Floor Houston, TX 77030	832.822.3180	jso@bcm.edu sstal@bcm.edu
Behavioral Issues	Dr. Kevin Krull	CCC, Meyer Center 6701 Fannin 16 th Floor Houston, TX 77030	832.822.3700	krkrull@texaschildrenshospital.org

ADULT CARE

SPECIALTY	DOCTOR	CLINIC ADDRESS	PHONE	EMAIL (if available)
Genetics	Dr. Maria Blazo, MD	The Baylor Clinic	832-822-4280	mblazo@bcm.edu

	Dr. Sharon Plon, MD, PhD	6620 Main, Ste. 1250 Houston, Texas 77030 6701 Fannin St. 16 th Floor Houston, Tx 7703		splon@bcm.edu
Neurology	Dr. Kinga Szigeti, MD	Department of Neurology Baylor College of Medicine 6501 Fannin St., NB302 Houston, TX 77030	713-798-5975	NA
Orthopedics	Dr. Heggeness, .MD Dr. Darrell Hanson, MD	Baylor Clinic 6620 Main, Ste. 1325 Houston, Texas 77030	713-986-6016	NA
Developmental pediatrics/learning disabilities	N/A			
Ophthalmology	Dr. Richard Lewis, MD	Alkek Eye Center Smith Tower, 6550 Fannin, Suites 1401 & 1501, Houston, Texas 77030	713-798-6100	rlewis@bcm.edu
Neurosurgery	Dr. Roc Chen, MD Dr. Hatem Megahed, MD	Department of Neurosurgery Baylor Clinic 6620 Main St. 13th Floor, Suite 1350 Houston, Texas 77030	713-798-4696	prchen@bcm.edu
Plastic surgery	Dr. Anthony Brissett, MD	6550 Fannin, Suite 1701 Houston, Texas 77030-3498	713-798-5900 713-441-3223	NA
Neuro-oncology	G. Rush Lynch, M.D.	6620 Main, 13 th floor Houston, Texas 77030	713-798-3750	lynch@bcm.tmc.edu
Medical Oncology/Radiation Oncology	G. Rush Lynch, M.D.	Baylor Clinic 6620 Main, Ste. 1375 Houston, Texas 77030	713-798-3750	lynch@bcm.tmc.edu
Endocrinology	Dr. Morali Sharma, MD	6620 Main, Ste. 1250 Houston, Texas 77030	713-798-4736	NA
Audiology/ENT	Dr. John Oghalai, MD	Bobby R. Alford Department of Otolaryngology - Head & Neck Surgery Smith Tower Suite 1701	713-798-5464	jso@bcm.edu

		6550 Fannin Street Houston, Texas 77030		
Radiology/ Neuroradiology	Dr. Susan Weathers, MD	Baylor Clinic Diagnostic Ctr. 6620 Main, Ste.1275 Houston, Texas 77030	713-798-2300	NA
General Surgery/Surgical Oncology	Dr.Charles Brunicardi, MD	Department of Surgery Baylor Clinic 6620 Main St. 14th Floor, Suite 1475 Houston, Texas 77030	713-798-5700	NA
Dermatology	Dr. Sylvia Hsu, MD Dr. Theodore Rosen,MD	6620 Main, Ste. 1425 Houston, Texas 77030	713-798-6131	NA
Cardiovascular Disease	Dr. Wayne Franklin, MD Dr. Douglas Mann, MD	The Baylor Heart Clinic 6620 Main, Ste. 1225 Houston, Texas 77030	713-798-2545	NA
Oral and Maxillofacial Surgery	Dr. Terry Taylor, DDS	6560 Fannin , Suite # 614 Houston, Texas 77030	713-790-1995	NA
Behavioral Issues	Baylor Psychiatry Clinic	Houston Medical Center Professional Building, 6655 Travis, Suite 500 Houston, Texas 77030	713-798-4857	NA

4. NUMBER OF NF PATIENTS YOUR CLINIC SEES

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

Insert numbers below

	NF1	NF2	SCHWANNOMATOSIS	OTHER
NUMBER OF PATIENTS SEEN IN PAST 12 MONTHS	96	6	0	24
NUMBER OF <u>NEW</u> PATIENTS SEEN IN PAST 12 MONTHS	*	*	*	*

TOTAL	96	6	0	24
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* We do not see new patients in our NF clinic. We require patients without a definite diagnosis to be seen in one of our other clinics (pediatric genetics, or adult genetics) first. Once a diagnosis is made/confirmed by the attending geneticist, then they are transferred to the NF Clinic.

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

Under 18 _____ 18+ _____ (give numbers - if data available)

OR estimate

Under 18 (%) 90% 18+ (%) 10%

5. TRANSITIONING PEDIATRIC TO ADULT NF CARE

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

One of the staff geneticists, Dr. Maria Blazo, is dual trained in Family Medicine and clinical genetics. Dr. Blazo has an active practice in Family Medicine at our Baylor Clinic location. As patients transition to adulthood, we provide Dr. Blazo's information if they choose to select her as their primary care physician. For all patients we continue to see patients as they enter adulthood in our clinic and will send letters and contact their adult PCP provider with any recommendations from our clinic.

6. INTERNAL CONFERENCES

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

The neuro-oncology team and tumor board meet separately on a weekly basis coordinated by the Texas Children's Cancer Center. Dr. Plon receives notification of all patients to be presented at those conferences. Either Dr. Plon or Dr. Blazo will attend the conference to aid in decision making for treatment decisions for NF patients. Patients with NF1 related tumors (optic glioma, JPA, MPNST) or NF2 (vestibular schwannoma, meningioma) may be presented at the appropriate team meeting.

7. CLINICAL TRIALS

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

Yes No

If 'no', briefly describe why.

Do you currently refer patients to clinical trials?

Yes No

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

In addition to directly participating in studies (such as the natural history study of plexiform neurofibromas) Dr. Plon refers appropriate patients to the medical oncologists of the Texas Children's Cancer Center, Drs. Susan Blaney, Alberto Pappo and Murali Chintagumpala. Dr. Blaney directs the Phase I clinical trials program and has participated in a number of treatment trials that including participation in the farnesyl transferase inhibitor study. Dr. Pappo and Dr. Chintagumpala have participated in treatment trials (using established agents) for solid tumors and brains tumors in NF1 patients.

8. PATIENT REGISTRY

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

Yes No

If 'yes', please describe. This is an IRB approved database/ patient registry that includes young patients with NF1 seen in the Neurofibromatosis clinic who are screened with neuro-imaging and/or ophthalmologic examinations per the guidelines for care of patients with NF1. This protocol is entitled: OPTIC GLIOMA IN NF1: SCREENING AND LONG-TERM FOLLOW-UP. Dr. Richard Lewis also has an ongoing database (over twenty-five years) of all patients (N>1000) that he has evaluated for ophthalmic manifestations of NF1.

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

× Yes *

No

*We would be willing to prospectively include patients in the centralized CTF NF Database assuming we obtain the appropriate IRB approval. The existing consent does not provide for transfer of data.

9. PUBLICATIONS and RESEARCH (IF APPLICABLE)

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

Airewele, G.E., Sigurdson, A.J., Wiley, K.J., Frieden, B.E., Calderera, L.W., Riccardi, V.M., Lewis, R.A., Chintagumpala, M.M., Ater, J.L., **Plon, S.E.**, and M.L. Bondy (2001). Neoplasms in Neurofibromatosis Type 1 are Related to Gender and Inheritance but not Family History of Cancer. *Genetic Epidemiology*, **20**:75-86.

Blazo, M .A. , Lewis, R.A. , Chintagumpala, M.M. , Frazier M. , McCluggage, C. and S.E. Plon (2004). Outcomes of Systematic Screening for Optic Pathway Tumors in Children with Neurofibromatosis Type I. *American J Medical Genetics*, *127A(3)*:224-9.

Plon, S.E., and V.M. Riccardi (1999). “The Phakomatoses and Other Neurocutaneous Syndromes” in Oski’s Pediatrics: Principles and Practice, 3rd Edition, (Eds. Frank A. Oski et al.) J. B. Lippincott Company, Philadelphia, 417: 2019 - 2025.

Plon, S.E. (2005). “The Phakomatoses and Other Neurocutaneous Syndromes” in Oski’s Pediatrics: Principles and Practice, 4th Edition, (Eds. Frank A. Oski et al.) J. B. Lippincott Company, Philadelphia.

Plon, S.E. and M.A. Blazo (2002). Neurofibromatosis I, in, *UpToDate in Pediatrics*. - updated annually

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

OPTIC GLIOMA IN NF1: SCREENING AND LONG-TERM FOLLOW-UP. This project will fulfill the following goals: 1. Analyze outcomes of patients screened both by systematic neuroimaging and regular ophthalmologic examinations according to established guidelines for the evaluation and management of children with NF1 at risk for OPT. This outcomes study aims to improve the care of patients with NF1 through an evidence-based approach to the detection of OPT. 2. Assess the impact of early detection on the reduction of morbidity of vision loss in children who are screened prospectively for OPT with ophthalmologic and neuroimaging examinations. 3. Facilitate future research and therapeutic intervention: Long-term follow-up of patients with NF1 will characterize increased incidence of second malignancies (particularly CNS) in NF1 patients with OPT; and identify patients that should be considered as candidates for any specific new treatment or intervention including preventative measures.

10. PATIENT SUPPORT

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

If 'yes' provide details.

Texas Neurofibromatosis Foundation

6555 Sierra Drive

Irving, Texas 75039

www.texasnf.org

Local Houston contact: Cindy Hahn, M.Ed., Managing Director, regularly attends monthly NF clinics in Houston. This organization also supports an annual patient/family educational symposium on NF (held at TCH this past May) and hosts a summer camp for children and families affected with NF.

If 'no', are you interested in starting such a group?

What resources would help you to do this?

11. OTHER INFORMATION

Please provide any additional information that is pertinent to your request to join the CTF NF Clinic Network.