

# Neurofibromatosis (NF): Defining Standards of Care

7:30am–4:45pm, Tuesday, April 28, 2009

450 Masonic Cancer Center Research Building

University of Minnesota

The schedule will be as follows:

8:00–8:30	Registration/Breakfast
8:30–9:30	Surgical issues: David Polly Jr., M.D.
9:30–10:30	Comprehensive visit for patients with NF: Elizabeth Siqveland, M.D., R.N., C.N.P.
10:30–10:40	Break
10:40–11:30	David Largaespada, Ph.D.
11:30–1:30	Lunch will be provided (Nancy Ratner, Ph.D. of Cincinnati Children's Hospital will be speaking from 12:00 – 1:00 in 450 MCRB)
1:30–2:30	Genetics: Susan Berry, M.D.
2:30–3:30	Neuropsychological Issues: Alicia Kunin-Batson, Ph.D.
3:30–3:40	Break (refreshments will be provided)
3:40–4:30	Oncologic issues and NF: Christopher Moertel, M.D.
4:30–4:45	Highlights and wrap up

Sponsored in partnership with: Children's Cancer Research Fund; University of Minnesota Medical School, Department of Pediatric Hematology/Oncology; The University of Minnesota Brain Tumor Program

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**Elizabeth Sigveland, M.S.N., R.N., C.N.P.**, is a certified pediatric nurse practitioner at Children's Hospitals and Clinics of Minnesota. At the Neurocutaneous Syndromes Clinic Without Walls, she provides casemanagement services, including initial patient evaluations. Sigveland has a master of science degree in nursing from the University of Minnesota.

**David Andrew Largaespada, Ph.D.** Dr. Largaespada is an authority on mouse genetics, gene modification and cancer genes. He received his B.S. in Genetics and Cell Biology from the University of Minnesota in 1987 and his Ph.D. in Molecular Biology with Dr. Rex Risser at the University of Wisconsin-Madison in 1992. He spent five years as a postdoctoral fellow in Frederick, Maryland at the National Cancer Institute working with world-renowned geneticists Dr. Nancy Jenkins and Dr. Neal Copeland, where the Leukemia and Lymphoma Society of America awarded him a postdoctoral fellowship. He joined the faculty of the University of Minnesota in late 1996. He is currently a Full Professor in the Department of Genetics, Cell Biology and Development and the Department of Pediatrics at the University of Minnesota. He is the leader of the Genetic Mechanisms of Cancer Program in the University of Minnesota Cancer Center and holds the Margaret Harvey Schering Land Grant Chair in Cancer Genetics. Dr. Largaespada was awarded the McKnight Land-Grant Professorship in 2000 and is an American Cancer Society Scholar. He has published over 80 scientific articles and teaches courses in the molecular biology of cancer and mammalian gene transfer. Dr. Largaespada has co-founded two biotechnology companies.

Dr. Largaespada's laboratory is working to exploit insertional mutagenesis for cancer gene discovery and functional genomics in the mouse. Mouse models of murine leukemia virus induced acute myeloid leukemia (AML) are being used to identify and characterize genes that have a role in leukemia progression after disease is initiated by mutations relevant to human AML. Ongoing work also includes genetic studies of myeloid leukemia chemotherapy resistance and relapse. The Largaespada lab has invested heavily in the use of a vertebrate-active transposon system, called Sleeping Beauty (SB), for insertional mutagenesis in mouse somatic and germline cells and for gene therapy. SB is being used as a tool for forward genetic screens for cancer genes involved in solid tumors such as sarcoma, prostate, gastro-intestinal tract and lung cancer.

**Susan Berry, M.D.** The major priority in Dr. Berry's laboratory is defining the molecular actions of growth hormone. She uses the model of the rat serpin multigene family, as several rat serpins are growth hormone responsive and developmentally expressed. As such, they are useful for investigation of the role(s) that hormones have in influencing the program of orderly gene expression in development. She developed the rat Spi 2.1 locus as a model system for the study of growth hormone response: this gene is now the most frequently used and best characterized model for the study of growth hormone action. Current projects include definition of the cis DNA element binding the GH inducible nuclear factor (GHINF) complex and determining the additional components of the GHINF complex, having demonstrated the participation of Stat5 in this signal transduction pathway.

In addition, the serpins respond to acute phase stimuli (inflammation). Several lines of evidence suggest there may be an interaction between this normal physiologic response to stress and growth regulation. Investigation of this interaction may provide new insights into the modulation of growth and the role of serpins in this modulation. In the rat Spi 2 locus there are both negative and positive acute phase reactants, while the promoters for these genes remain highly homologous. This permits a detailed examination of the interaction of both cis and trans factors

in the coordinate regulation of the positive and negative responses to inflammation. Her current project in this area is defining the role of STAT proteins in the response to inflammation, using the rat Spi 2 gene model.

An additional area of investigation in the lab is to define molecular mechanisms by which gene expression can be altered during maturation, and to evaluate the participation of hormonal signals in development. Prenatal growth may be mediated by different hormonal stimuli than growth after birth. Work is ongoing to define the participation of GH in perinatal growth and gene expression, currently investigating a relative resistance to growth hormone action in perinatal life.

**Alicia Kunin-Batson, Ph.D.** is a pediatric neuropsychologist and an assistant professor in the Division of Pediatric Clinical Neuroscience in the Department of Pediatrics. She received her bachelor's degree in psychology from Kenyon College in Gambier Ohio and attended Finch University of Health Sciences/The Chicago Medical School for graduate training in clinical psychology, where she received her M.S. degree in 1997 and Ph.D. degree in 2001. Dr. Kunin-Batson completed internship and fellowship training in pediatric psychology at the Children's Hospital of Philadelphia, and fellowship training in pediatric neuropsychology at the University of Minnesota.

Dr. Kunin-Batson provides psychological consultation to cancer survivors and their families in the University's Long Term Follow-Up Clinic, and conducts neuropsychological evaluations of children, adolescents, and young adults to assess for potential late effects of cancer treatments on learning.

**Christopher Moertel, M.D.** Dr. Moertel graduated from the University of Minnesota Medical School. He completed his internship and residency in Pediatrics at the Baylor College of Medicine in Houston, Texas. He then completed his fellowship in Pediatric Hematology and Oncology at the Mayo Clinic/ Mayo Graduate School of Medicine in Rochester, Minnesota. He joined the medical staff at Children's Hospitals and Clinics of Minnesota in 1990. While at Children's, he served as Chief of Staff, Medical Director of Hematology/Oncology and Lead Physician in the Theodora Lang Oncology Clinic, among other leadership positions. While at Children's, he was co-director of neuro-oncology, founder and medical director of the Neurocutaneous Syndromes Clinic Without Walls, and attending physician in pediatrics at the Fairview/University Hemophilia and Thrombosis Center. Dr. Moertel has served on numerous national and local professional committees and is the author of a number of book chapters, articles and abstracts. Special interests include rare pediatric tumors, neurofibromatosis-associated neoplasia, and the therapy of children with brain and spinal cord tumors.