1999-2000
Summary of Activities
USA Comprehensive Sickle Cell Center

General Operations: We have consolidated our Sickle Cell Center Administrative Office and Research Laboratories into the second floor of the MSB. The Sickle Cell Center Administrative Offices are located in MSB 2015. The laboratories of Dr. Steven Goodman, Dr. Betty Pace, Dr. Johnson Haynes, Dr. Gan Wang and Dr. Mohan Bhatnagar are now located in the second floor MSB expansion.

Research: We were well-represented at the 24th Annual National Sickle Cell Program meeting held in Philadelphia, PA, in April 2000. Our faculty, students, and staff who presented and their subjects were: "NAC Blocks Dense Cell and ISC Formation In Vitro by Protecting the Gardos Cannel and β-Actin from Oxidative Damage," Dr. Archil Shartava; "WEB 2170 and Zileuton Inhibit Enhanced Sickle Red Blood Cell (SRBC) Retention Mediated by the Activated Polymorphonuclear Cells (A-PMN) in the Lung Circulation," Dr. Johnson Haynes; "Development of a DNA Vaccine for Human Parvovirus B19," Dr. David Dean; "Molecular Mechanism of Peptide Nucleic Acids (PNA) Binding-inducted Gene Expression," Dr. Gan Wang; "Triplex DNA Formation Is Enhanced by Ethanol," Dr. Mohan Bhatnagar; "γGlobin Induction by a Novel Fetal Stage Specific Transcription Factor," Amy Ferry; "Fetal Hemoglobin Induction: Characterization of DNA-binding Proteins Altered by Butyrate," Curtis Browning; "Cytokines and Acute Phase Reactant Proteins in Acute Chest Syndrome, Vaso-occlusive Crisis and Steady State," Dr. Felicia Wilson; "Abnormalities of Growth and Pulmonary Function in Acute Chest Syndrome," Dr. Felicia Wilson.

Adult Operations: The 1999-2000 academic year marks the close of the second year that the Adult Sickle Cell Clinic has operated as a subspecialty clinic. The primary objectives of this clinic are (1) to be a resource for practicing physicians in the University and private community; and (2) to serve as a positive educational experience for primary care residents in the Departments of Medicine and Family Medicine at the University of South Alabama. The clinic is staffed by Dr. Johnson Haynes, Jr., and Ms. Ardie Pack-Mabien, Nurse Practitioner. Over the last academic year, 32 residents have rotated through the outpatient Adult Sickle Cell Clinic. The Adult Sickle Cell Clinic continues to serve the greater Mobile area in addition to a continuous influx of patients from Thomasville, Bay Minette, Mt. Vernon, Evergreen, and Montgomery, AL. In addition, we are seeing a small number of patients from Pensacola and Panama City, FL, and Pascagoula, MS. All referring physicians are written correspondence outlining the care received in the adult clinic as well as the planned therapeutic course implemented for that patient's condition. Ms. Pack-Mabien continues to support the pediatric outpatient clinical operations and serves as the liaison when transitioning patients from the pediatric to the adult services. There continues to be a gap in transitioning patients who are on chronic transfusion therapy which require iron chelation for iron overload.

The challenges for the future in the adult clinical operation continues to be (1) establishment of an Ambulatory Pain Clinic geared towards managing pain which exceeds home management, but may not necessitate hospitalization; (2) to be creative in the development and participation of the adult hematology services in caring for adults requiring chronic transfusion therapy and desferal chelation; and (3) the continued development of strong clinical projects in caring for patients with sickle cell disease.

Clinical investigational projects currently underway at the University of South Alabama Comprehensive Sickle Cell Center inclusive are: (1) The use of the fentanyl patch in managing painful vaso-occlusive crisis; (2) n-acetylcysteine, a potential therapeutic agent in the management of vaso-occlusive crisis; (3) zileuton, a potential therapeutic agent in the management of vaso-occlusive crisis and the acute chest syndrome; and (4) the role of cytokines in acute chest in sickle cell patients.

Pediatric Programs: Dr. Betty Pace serves as the Associate Director for Pediatric Programs, working in conjunction with Drs. Felicia Little-Wilson and Yang to provide comprehensive medical care and services to pediatric sickle cell patients. A major goal of the program has been to establish a separate sickle cell clinic, independent of the general hematology clinic. The number of patients continues to grow for both services, making it a necessity to have a second clinic. Negotiations are ongoing with the Department of Pediatrics to achieve this goal. Our clinical staff consists of Ms. Pack-Mabien, a licensed Nurse Practitioner who serves as the Nurse Coordinator for Clinical Programs and cares for patients in the pediatric and adult clinics. Ms. Stephanie Durggin serves as the Pediatric Nurse Coordinator developing protocols for patient medical care and education. Ms. Evelyn Bright is the sickle cell nurse whose primary responsibilities are to facilitate ongoing clinical trials and projects and to assist with clinic operations. A second major focus in the pediatric program has been the establishment of a Children Youth Sickle Network to extend medical care to sickle cell patients in the nine counties we serve, by providing on-site services in the rural communities. A Telemedicine approach will be utilized to staff these satellite clinics. We are continuing efforts to establish an Ambulatory Pain Clinic, to be co-directed by Drs. Pace and Haynes.

Psycho-Social Research Division: The primary goal of the Psycho-Social Research Division (PSRD) this year has been to develop the framework of a new arm within the Comprehensive Sickle Cell Center. This new arm will encompass collaborative efforts with several other departments within the University, the Community Sickle Cell Program, and Franklin Primary Health Centers. The division was successful in garnering support from Dr. Steven Picou, Chair of Sociology and Anthropology, to allow two of his staff members, James Lee, Ph.D., and Nicole Flynn, Ph.D., to serve on the division's technical advisory committee. Elise Labbe, Ph.D., from the Psychology department, Dr. Arvind Shah, Mathematics department, Linda Jones, Executive Director of SCDAA-Mobile Chapter, and a representative from Franklin Primary Health Center will also serve on this committee. This committee is committed to developing a strong proposal for the year 2003 Sickle Cell Center renewal process. The PSRD will continue to build on the progress started this year when we conducted an open house with over three hundred people attending, and

developed an adult support group, who hold their meetings on the USA campus. PSRD staff assisted the clinical research division in the recruiting of patients for various projects. PSRD developed the first USA Comprehensive Sickle Cell Center newsletter. PSRD is collaborating with the National Sickle Cell Disease Association on several projects to strengthen their member organizations program services while using survey instruments developed by the PSRD and the Statistical Core here in Mobile. Our goal is to have their member organizations across the country help us to standardize data collection survey instruments designed by the USA Sickle Cell Center over the last ten years. The PSRD has also strengthened the lines of communication with the local community-based program for sickle cell services by providing ongoing training for staff involved in the Center grant and technical assistance with the various Center-related projects.

Education: The Center faculty continue to provide a broad series of lectures throughout the academic and private medical community. The Center faculty continues to support the Educator/Counselor Certification Program which took place three times this academic year at the Sickle Cell Disease Association of America, Mobile Chapter. Ms. Ardie Pack-Mabien, Nurse Practitioner, has given fourteen lectures over the last academic year to nurses within the University of South Alabama, private hospitals in the Mobile community, School of Nursing, and to outlying nursing personnel in the surrounding counties. The Center faculty (Drs. S. Goodman, Dr. J. Haynes, Dr. B. Pace, A. Pack-Mabien and R. Peterson) developed and conducted two symposia on "Practical Issues in Sickle Cell Disease, Clinical and Psycho-social" in Montgomery and Huntsville, AL. The faculty continues to participate in the National Sickle Cell Disease Program which was held in Philadelphia, PA, in April of 2000. Ongoing basic research was presented on laboratory investigation conducted in the labs of Drs. Goodman, Haynes, and Pace. In addition, Dr. Felicia Wilson presented data collected from her clinical research projects related to pulmonary function abnormalities associated with the acute chest syndrome. The Summer Student Research Program continued this academic year. Jessie Knight participated in an ongoing collaborative project between Drs. Haynes and Pace investigating potential therapeutic effects of the 5-lipoxygenase enzyme inhibitor and analog of hydroxyurea, zileuton, in the management of acute chest syndrome. John Burch, USA medical student, worked in Dr. Pace's laboratory on a project entitled "Characterization of DNA-binding Proteins Altered by Butyrate in the Globin Promoter." The survey on Nurses' Attitudes and Practices in the Treatment of Acute Pain Episodes has been revised and submitted as of September 2000 to Applied Nursing Research. The Physicians' Attitudes and Practices survey data have been collected from 8 of 10 of the NIH-funded Sickle Cell Centers, and the analysis of this data has been completed. Through these continued efforts, such educational support will lead to a stronger health care delivery system.