

**2000-2001  
Summary of Activities  
USA Comprehensive Sickle Cell Center**

General Operations: As of August 1, 2001, Dr. Johnson Haynes, Jr., replaced Dr. Steven Goodman as Director of the USA Comprehensive Sickle Cell Center. The Sickle Cell Administrative Office and Research Laboratories are located on the second floor of the MSB. The Sickle Cell Center Administrative Offices are found in MSB 2015. The laboratories of Dr. Betty Pace, Dr. Johnson Haynes, and Dr. Mohan Bhatnagar are located in the second floor MSB expansion. The sickle cell Laboratory Core and office of the Children Youth Sickle Network are now located in the second floor expansion of MSB.

Research: We were well-represented at the 25<sup>th</sup> Annual Sickle Cell Program meeting in New York, NY, on April 17-19, 2001. Faculty, students, and staff who presented were: Dr. Johnson Haynes, Jr. (Zileuton: An Inhibitor of Activated Polymorphonuclear Cell Mediated Sickle Erythrocyte Retention in Lung and Promoter of Hemoglobin F Induction *In vitro*); Dr. Steven Goodman (Erythrocyte Spectrin Is an E2 Ubiquitin Conjugating Enzyme That Is Blocked in Sickle Cell); Dr. Solomon Ofori-Acquah (The Role of Antisense Globin Transcripts in Gene Therapy of Sickle Cell Disease); Heather Foley (Globin Gene Regulation by the JAK/STAT Signal Transduction Pathway); Dr. Mohan Bhatnagar (Neutrophil Chromosomal Domain in Sickle Cell Disease); Dr. Gan Wang (The D-Loop Length Requirement for Peptide Nucleic Acids Binding Indirect Transcriptions); and Ms. Betty Robinson (Southwest Alabama Children Youth Network: Telemedicine as a Model for Health Care Delivery).

Current projects in the USA-CSCC are Novel Mechanisms of Globin Gene Silencing During Alternate Lineage Commitment, Betty Pace, PI; Modulation of Globin Expression by Antisense Transcripts (Scholars Project), Solomon Ofori-Acquah, PI; Mechanisms of Fetal Hemoglobin Induction, Danna Zimmer, PI; and Acute Lung Injury in Sickle Cell Disease, Johnson Haynes, Jr., PI.

Adult Operations: The 2000-2001 academic year marks the close of the third year that the Adult Sickle Cell Clinic has operated as a subspecialty clinic. The primary objectives of this clinic are to (1) 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, 17 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 surrounding counties. In addition, we are seeing a small number of patients from Pensacola and Panama City, Florida, and Pascagoula, Mississippi. 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 and require iron chelation for iron overload.

The principal challenges for the future in the adult clinical operation continues to be the development and participation of the adult hematology services in caring for adults requiring chronic transfusion therapy and desferal chelation.

Clinical investigational projects currently underway at the University of South Alabama Comprehensive Sickle Cell Center inclusive are (1) N-acetylcysteine, a Potential Therapeutic Agent in the Management of Vaso-occlusive Crisis; (2) Zileuton, a Potential Therapeutic Agent in the Management of Vaso-occlusive Crisis and the Acute Chest Syndrome; and (3) The Role of Cytokines in Acute Chest Syndrome in Sickle Cell Patients.

Pediatric Programs: Dr. Betty Pace serves as the Associate Director of Pediatric Programs, working in collaboration with Drs. Wilson and Yang to provide comprehensive medical services to pediatric sickle cell patients. A major goal of the pediatric program over the last year has been the establishment of a Children Youth Sickle Network (CYSN) to serve clients in the eight counties we serve in rural Alabama. Funding has been secured through the Robert Wood Foundation with matching funds from the Dean's office; satellite clinics will be initiated this fall. A telemedicine approach will be used to accomplish this unique program in the College of Medicine. Delivering some of the medical services in satellite clinics will aid in solving the problem of high patient census in the weekly sickle cell clinics held in Mobile. We will continue to work towards expanding the number of clinic days so that adequate medical services can be delivered to patients in a timely fashion. Clinic protocols were developed for the competitive renewal of the Comprehensive Sickle Cell Center grant application. During the upcoming year we plan to develop two new clinical protocols to secure salary support for the hematology/oncology faculty and research nurse. Other areas of clinical care will be examined to improve the transition from pediatric to adult services.

Psycho-Social Research Division: The PSRD directed by Rosie Peterson, serves as the arm of the USA Comprehensive Sickle Cell Center that assures the behavioral, cognitive, and psycho-social programs are integrated in the overall care plan of individuals with SCD. Programs within the PSRD focus on the prediction of medical compliance, access to healthcare, quality of treatment, and overall psycho-social functioning of the families and children with SCD. PSRD also strives to influence the overall healthcare delivery system for individuals with sickle cell disease in the catchment area served by our Center. PSRD participated in the following activities during 2000-2001: Nationwide Survey: a simple, one-page service inventory survey was developed and mailed to all SCDA member organizations to identify the types and extent of services being provided by member organizations. Out of 62 SCDA member organizations, 36 organizations responded to the questionnaire. The questionnaire yielded the following profile of SCDA member organizations: the majority of organizations (78%) provide both the general and the

professional education. Of those organizations which provide only one type of education, more organizations provided only professional education. However, more general educational events were held than professional educational events. Approximately 56% of SCDAA member organizations held 900 tutorial programs. The number of participants ranged from 4 to 60 with the exception of one, which had 118 participants. Almost all of SCDAA member organizations provide sickle cell screening; a majority of them provides mass and individual screening. Congressional Hearing on Universal Health Care: The PSRD director was asked to write a statement for the hearing in support of the needs off universal healthcare for every American, as well as to highlight ongoing issues created by existing limitation in access to health care for our consumers with sickle cell disease. Educational Series of Pre- and Post-Test Survey Instruments: An educational series of pre- and post-test survey instruments were developed as scannable teleforms and tested by the Sickle Cell Disease Association in Mobile. Since these forms were developed by the USA Sickle Cell Center with NIH funding over the last ten years, the plan is to standardize and copyright these forms for usage on a national level. Medicaid Project: The purpose of this project is to create discussion and a solution to the 16-day limit on inpatient hospital stays for adults who are currently receiving Medicaid and the ongoing issues this limitation creates for our adult Alabamians with sickle cell disease. Initiatives for Children and Adolescence Research Center: PSRD, along with Dr. James and Dr. Elise Labbe developed a plan to create a center, called the Center for Childhood and Adolescence Research. This center will address experiences across multiple social institutions that promote or hinder cognitive development and learning among youth. The intent is for our center to promote collaborative, interdisciplinary research that will evaluate children's involvement in important institutions (such as family, healthcare, and schools) and the conditions that prevent some from gaining benefits from full participation. A grant for this center is pending.

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 eight 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 nursing personnel in the outlying, surrounding counties. The faculty continues to participate in the National Sickle Cell Disease Program, which was held in New York, NY, in April 2001. Kristi Reese, 2<sup>nd</sup> year medical student, worked in the laboratory of Dr. Betty Pace on the "Involvement of the ERK MAP Kinase Pathway in  $\gamma$  Globin Gene Expression." The work was presented at the 28<sup>th</sup> Annual Medical Student Research Day. The USA-CSSC co-sponsored the speaker for the 28<sup>th</sup> Annual Medical Student Research Day, Dr. Robert Hebbel, whose topic was "Endothelial Biology of Sickle Cell Disease." The survey on Nurses' Attitudes and Practices in the Treatment of Acute Pain Episodes has been accepted to the *Journal of Applied Nursing Research*. The Physicians' Attitudes and Practices survey has been completed and analyzed and is in preparation for journal submission. Through these continued efforts, such education support will lead to a stronger health care delivery system.

### SUMMARY OF BIOTECHNICAL SERVICES

Biotechnical Services consist of the Biopolymer, Research Cytometry, Mass Spectrometry and Transgenic Animal/ES Cell core laboratories and report to the Office of the Senior Associate Dean. Each biotechnical core laboratory is managed by a director with input from a core advisory committee. The chart shown below outlines the organizational structure of the Office of Biotechnical Services for the 2000-2001 academic year. The following are directors for each core laboratory: Dr. Tin Cao, Director of the Biopolymer Laboratory; Dr. Raymond Hester, Director of the Research Cytometry Laboratory, Dr. Ann Abraham, Acting Director of the Mass Spectrometry Laboratory; Drs. Danna and Warren Zimmer, Co-directors of the Transgenic Animal/ES Cell Laboratory.

2000-2001  
 University of South Alabama  
 College of Medicine  
 Organizational Structure  
 Office of Biotechnical Services

