HANDOUT #1

Arkansas Legislative Task Force
On Sickle Cell Disease

REPORT TO THE ARKANSAS GENERAL ASSEMBLY

August 31, 2010

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Executive Summary

Statement of the Problem

Sickle cell disease is an inherited blood disorder that affects over 1,000 persons of all races in Arkansas. Persons affected by sickle cell disease have red blood cells that tend to become hard and sticky. These changed cells tend to stick together in small vessels, blocking the blood flow, which leads to intense pain, anemia, and organ damage in the affected area. Common complications include anemia, acute chest syndrome, stroke, skin ulcers, and infections.

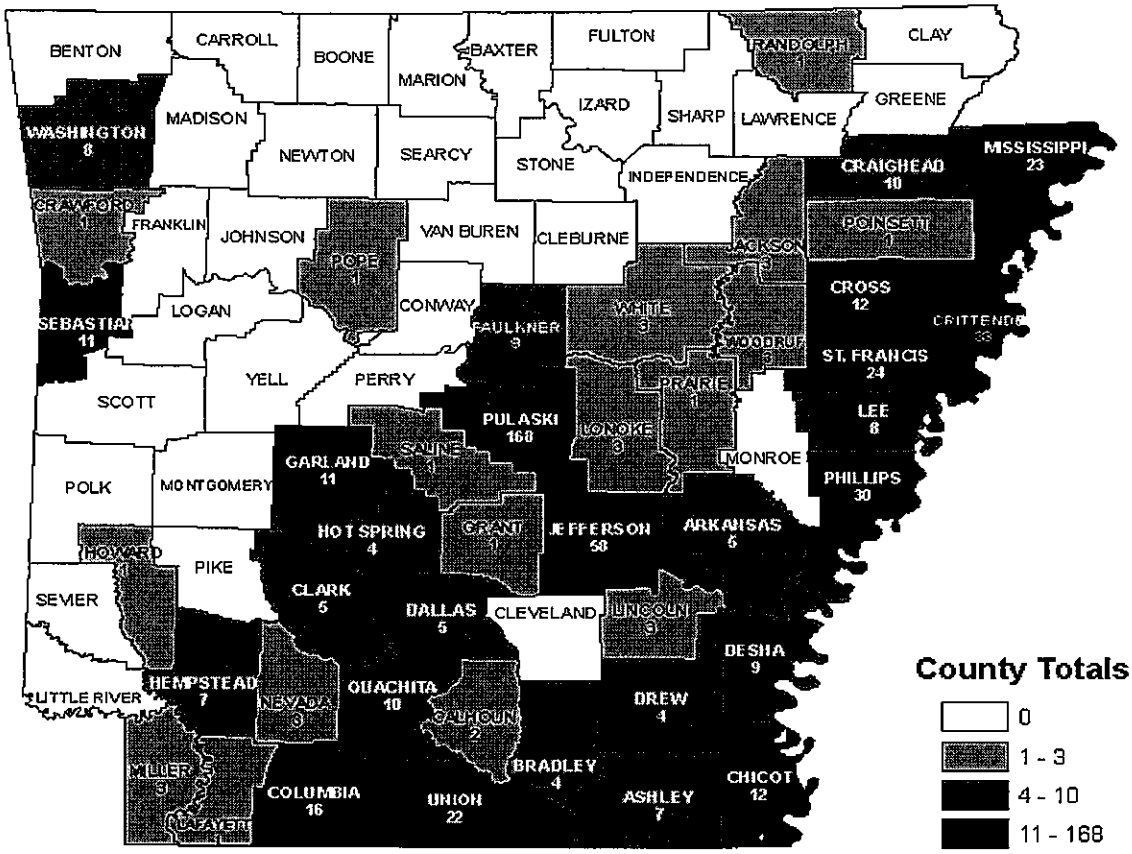
Only a couple of decades ago, most children with sickle cell did not reach adulthood. Because of that fact, there were few doctors who treated adults. Even though the life expectancy has greatly increased, adult services have not caught up with these changes. There is no self-identified clinic in Arkansas for adults with sickle cell disease. Because of poor quality of care, adult patients are now placing a large demand on the health care system. Few adult providers understand how to care for individuals with sickle cell disease. With poor quality care, many adults with sickle cell disease have frequent pain episodes that result in misery and poor health outcomes. Because of their poor health, many people with sickle cell are unemployed. Their economic status means that a person with sickle cell may be uninsured. With frequent emergency department visits and hospital admissions, hospitals are often faced with unreimbursed care. Persons unable to work because of poor health are also unable to pay taxes. It is advantageous to the patient, hospital, and society to improve health outcomes for individuals with sickle cell disease. Establishment of an adult clinic can improve health outcomes for patients and provide cost savings to patients, hospitals, insurers, and through Medicaid savings, the state and federal government.

Key Recommendations

- Develop a Comprehensive Sickle Cell Program, in which children and adults can receive state of the art care and case management, using central centers and peripheral sites.
- Allow Medicaid payments for more than four medications per month and six hospital days per year. This should result in better health outcomes for patients.
- An educational outreach coordinator should be hired to help the agencies that provide educational services integrate their activities and to address unmet training needs.
- Create a sickle cell disease registry within the Arkansas Department of Health or the University of Arkansas for Medical Sciences, with mandatory reporting.
- Fund a team of research assistants to develop pilot studies about sickle cell disease.
- Conduct a general awareness campaign using television, radio, and newspaper advertising.
- Target physicians using direct mail and conferences for family practice physicians and pediatricians.
- Use schools to reach pre-teens for education about sickle cell trait and its implications for their lives.

Total Infants with Sickle Disorders (S/S, S/C, S-β-thal) Detected through Newborn Screening

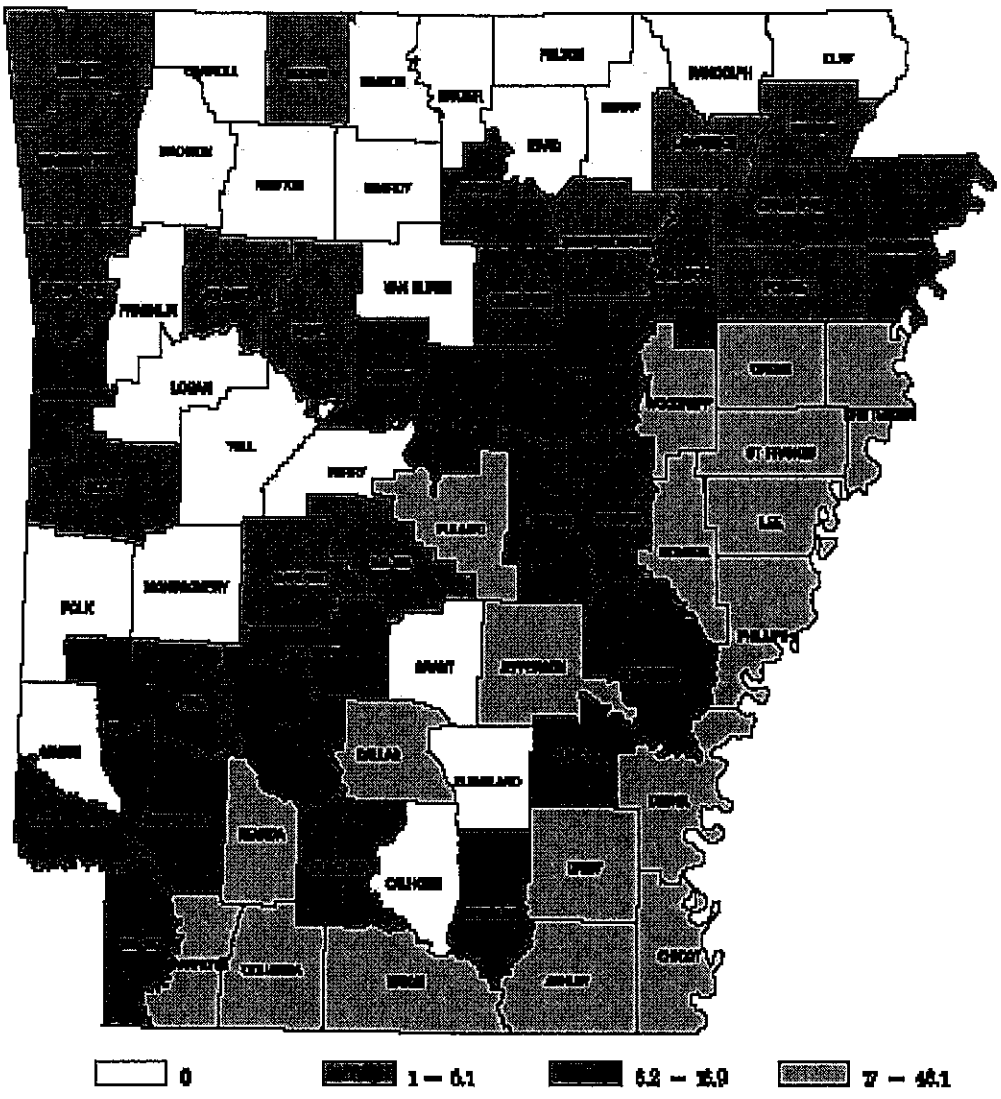
Arkansas,* October 1988 - September 2009 by County of Residence



*Includes Arkansas resident infants detected through adjoining states' newborn screening programs

Source: Arkansas Department of Health, Newborn Screening Program

All Sickle-cell Patients with any type of claim during SFY 2008
Prevalence per 10,000 Beneficiaries [0-64] by County of Beneficiary



Statement of the Problem

Sickle cell disease is an inherited blood disorder that affects over 1,000 persons in Arkansas. Persons affected by sickle cell disease experience structural changes in their hemoglobin, which causes red blood cells to become hard and sticky. These changed cells tend to stick together in small blood vessels, blocking the flow of blood. The obstructed blood flow leads to oxygen and nutrients deficits in various tissues, resulting in intense pain, anemia, and organ damage in the affected area. Common complications for various individuals include splenic sequestration, aplastic crisis, acute chest syndrome, stroke, skin ulcers, and severe infection.

Only a couple of decades ago, most children with sickle cell did not reach adulthood. With improved treatment, the life expectancy of a person with sickle cell disease has more than doubled in just a couple of generations. As a result of the increased life expectancy, adult patients are now placing a large demand on the health care system. Few adult providers understand how to care for individuals with sickle cell disease. With poor quality care, many adults with sickle cell disease have frequent pain episodes that result in misery and poor health outcomes. Because of their poor health, many people with sickle cell are unemployed or underemployed. Their economic status means that often a person with sickle cell will be uninsured. With frequent emergency department visits and hospital admissions, hospitals are often faced with unreimbursed care. Persons unable to work because of poor health are also unable to pay taxes. It is advantageous to the patient, hospital, and society to improve health outcomes for individuals with sickle cell disease.

Treatment for persons with sickle cell disease has improved dramatically over the past 15 years. Since it is a low-incidence condition, many physicians are unable to keep up with the advances in care. With the complexity of the disorder, it is important for the patient to receive care from a knowledgeable specialist. Management of sickle cell pain is especially challenging. For pain from other causes, the typical practice is to start with a relatively small dose of medication and titrate up until the pain is controlled. With pain from sickle cell disease, that approach typically leaves the patient in pain for hours or even days. With this condition, the recommended protocol is to deliver a bolus (large dose) of medication to get ahead of the pain curve. Many health care practitioners misinterpret the continued requests for pain medication as drug seeking behavior and resist delivering an appropriate dose of medication. The inability to get their pain under control may lead the patient to feel disrespected by their physician and to become distrustful of the health care system in general.

While there are pockets of quality care in the state, there currently is no self-identified, adult clinic for persons with sickle cell disease. Establishment of such a clinic can improve health outcomes for the patient and cost savings to patients, hospitals, insurers, and through Medicaid cost savings, the state and federal government.

The Legislative Task Force on Sickle Cell Disease examined the conditions for persons with sickle cell disease and sickle cell trait, health care providers, and insurers. Their findings and recommendations are reported in this document.

Clinical Services

The care of sickle cell patients is fragmented, especially for adults. Although there is a comprehensive sickle cell center for children, at Arkansas Children's Hospital, there is no comparable adult program. Thus, adults (age 21 and older) have no "medical home" to transition to after they leave the pediatric program. Medical care designed to reduce morbidity and prevent crises in adults (and to a lesser degree in children) is not uniformly available throughout the state. Most adult patients are not receiving appropriate medication to improve their disease status (hydroxyurea), and are not being taught how to manage their pain in a way to prevent hospital stays. Medical providers are either unwilling or unable (because of lack of training or lack of support and guidance from an adult comprehensive program) to provide acute and chronic care to sickle cell patients of all ages, but especially adults, in their local community. Medicaid data suggest that adults with sickle cell disease visit their emergency rooms frequently and are hospitalized frequently because of their lack of a personalized plan of care. Additionally, some Medicaid rules and regulations actually make it more difficult for adults with sickle cell disease to receive the ambulatory care they need.

If these areas of deficiency are corrected, it could prevent or minimize morbidities associated with the disease, reduce hospitalizations, reduce medical costs, and in general improve the quality of life of affected persons.

The cost of establishing the recommended clinics is $696,875 for the first year and about $25,000 less in the following years.

Sickle Cell Disease Education

Education is an essential component of a plan to improve sickle cell outcomes. There is a need to increase the knowledge of professionals, families, and the general public. As patients gain better understanding of ways to manage their sickle cell disease through lifestyle choices they will have fewer pain episodes. It is particularly important for patients to understand how proper hydration, diet, rest, and responding to extreme heat and cold can help them avoid vaso-occlusive (pain) episodes. As family members become better informed, they will be able to support individuals with sickle cell disease. Persons with sickle cell trait need to understand the possibility of their children being born with sickle cell disease. School personnel need training to understand the unique educational needs of students with sickle cell disease, particularly the likelihood that those students may have frequent absences from school and may require assistance to keep up with their schoolwork during those absences. They should also receive training to be able to recognize the early stages of a pain episode and what to do in case of a low fever. Health care providers need access to education about the latest developments in treatment of sickle cell disease and will benefit from just-in-time information about specific cases.

There are four major steps involved in implementing a successful statewide Sickle Cell Disease Educational Plan:

- Understand the different communities within our state and how they receive information
- Prepare an official Statewide Sickle Cell Disease Educational Plan
- Establish community Sickle Cell Disease Educational Plans
- Evaluate Sickle Cell Disease Educational Plans to determine if they are working

Partnerships with organizations such as Community Health Centers and AHEC's, who are present in every region of the state and who are designed to expand advocacy and education, will play key roles in building a strong education program. Core Legislative Task Force members including Arkansas Department of Health (ADH), Arkansas Minority Health Commission (AMHC), University of Arkansas for Medical Sciences (UAMS), Arkansas Foundation for Medical Care (AFMC), Arkansas Children Hospital (ACH), Sickle Cell Support Services, and Mutual Sickle Cell Support Group should collaborate to deliver enhanced sickle cell disease education services statewide. The member agencies are engaged in education activities for professionals, patients, family members, and members of the general public as a part of their ongoing work. These agencies should work more closely to coordinate their efforts.

In order to coordinate these efforts and to address gaps in educational efforts the state will benefit from hiring a sickle cell disease educational outreach coordinator. This person could organize quarterly training events for professionals using the telehealth network to link remote communities through compressed interactive video and could plan presentations for professional conferences. The outreach coordinator could also provide training to a network of lay educators who could then take the lead in helping the general public gain a better understanding about sickle cell disease. The coordinator could assist in the education of persons with sickle cell disease by arranging speakers to present at support groups around the state and assist with an annual sickle cell conference. The cost of employing a coordinator and supporting the education plan is $91,750.

Supportive Services

Persons with sickle cell disease expressed a need for a wide array of supportive services. The comprehensive care clinic for children and youth at Arkansas Children's Hospital helps connect children to the services that exist. However, for adults fewer services exist and adult patients typically have little assistance in finding the few services that exist. The major source of such guidance comes from support groups in Little Rock, Pine Bluff, Helena, Monticello, and Clarksville. As useful as those resources are, the need for case management far outstrips their ability to assist. A major expansion of case management services and support groups is needed.

Unemployment or underemployment for adults with sickle cell leads to numerous other challenges because of lack of financial resources. Potential employers need to understand the condition and the need for accommodations, particularly the pattern of absences. Persons with sickle cell need assistance in finding jobs that are compatible with their health needs. A couple of needs that are often cited by persons with sickle cell disease are related to inadequate employment. The support groups in the state receive numerous requests for assistance with an array of financial needs. Those community-based organizations have resources to assist with only a small fraction of the requests they receive. A second need associated with limited employment is that of insurance. Since our insurance system is largely employment-based, many people with sickle cell disease are not insured. This results in many persons not receiving ongoing health care management, which could reduce the number of acute episodes that frequently result in the patient seeking care in the emergency department of the hospital. It also results in a sharp increase in the number of hospital admissions. Without insurance, the hospital encounters are often unreimbursed, resulting in financial strain for the hospitals. For a number of rural hospitals in the state, this burden puts them at risk of being unable to continue their survival. As is the case with other chronic conditions, persons with sickle cell disease may need supportive counseling or therapy. Often they find their therapists do not understand their disease process, which makes it difficult for the patient to find the help they need. Many also have difficulty paying for those services. This combination causes some people with sickle cell to forego the help they need. Students with sickle cell disease may experience frequent absences from school and need assistance with staying abreast of their coursework. Without this assistance, it is common for students to fall behind and be forced to repeat grades.

A persistent problem for adolescents is making the transition to adulthood. Many social services that exist for children and youth are no longer available for adults. Finding a physician who serves adults and is knowledgeable about sickle cell is a challenge. Further complicating that process is the need to prepare the adolescent to be responsible for his or her own health care. It is common for the child to depend on the parent to make all the health care decisions. When the person becomes an adult and is responsible for all their own choices, they may lack understanding about what they need to do to care for themselves. If they are not supported by a knowledgeable physician, they are less likely to make healthy choices.

The cost of supportive services is included in the budget for clinical services.

Sickle Cell Disease Registry

Need for and Potential Uses of a Registry

The true number of persons in Arkansas affected by sickle cell disease (SCD) is unknown at present. Estimates have been calculated based on those diagnosed as a result of newborn screening, as well as on hospital discharge and death certificate information. However, SCD is not always listed as a contributing cause for admission to a hospital or for death, even when it was present and did contribute. Some individuals with disease may not require medical care very often, and some may ultimately die of causes truly unrelated to sickle cell disease. For these reasons, current estimates of affected individuals in Arkansas may be low.

Creation of a comprehensive sickle cell registry would allow for a complete "census" of persons with SCD in Arkansas. Through collection of appropriate information, much knowledge could be gained about health status of individuals with SCD as well as the system of care for those individuals. The registry could collect data such as address, form of the disease, health insurance coverage, educational attainment, employment status, and receipt of disability assistance. In addition to basic demographic information, the registry could collect valuable clinical information. For example, information could be gathered to determine how many of those with SCD are receiving care in accordance with national guidelines, hospitalizations, health complications, immunization status, use of prophylactic antibiotics and folic acid regimens, and use of various therapies, such as hydroxyurea.

Once a registry is established and the above pieces of information are systematically assimilated and analyzed, a much clearer picture of sickle cell disease in Arkansas should emerge. Needs that before were ill-defined or intangible will become much better delineated, allowing for targeting of resources to the most urgent priorities. In particular, the plight of adults with SCD should become much more evident, which in turn should spur improvements in the system of care.

In addition to facilitating analyses of various characteristics of Arkansans with SCD and the system of care overall, creation of a registry would also allow for the possibility of recruitment of patients into clinical trials.

Mandatory Reporting and Location of Registry

To achieve the full benefit to public health, all individuals with disease must be included in the registry. Therefore, the Task Force recommends mandatory reporting of sickle cell disease diagnosis and individual health status information through specific authorizing legislation. Providers mandated to report should include the Newborn Screening Program (within the Department of Health), physicians, laboratories, and hospitals. Both the Arkansas Department of Health and the University of Arkansas for Medical Sciences (UAMS) have experience in maintaining disease registries and either agency could serve as the home for a registry.

The cost of establishing a registry is $174,665.

Research

A program of clinical care and educational services will be supported by a strong program of research and program evaluation. Trained evaluators are able to determine the efficacy of various treatments, examine the effectiveness of service delivery systems, identify social service needs and resources, and validate educational approaches. In order to improve outcomes for individuals with sickle cell disease and their families, it is essential to establish a research infrastructure in the state. While there are a number of well qualified researchers in the state, there has been little investigation about sickle cell. The primary research activities have taken place at Arkansas Children's Hospital, where the clinical team has been part of several evaluations of best practice in clinical care. The paucity of activities involving sickle cell disease has resulted in a lack of data that can be used to answer questions about clinical care and services.

New research ventures may be fostered by developing a research collaborative involving persons interested in developing new knowledge about sickle cell disease. Most of the agencies involved in the Task Force have a vested interest in learning more about the condition and could form the nucleus of such a collaborative. Other agencies and hospitals should be invited to join the effort.

Establishing an adult clinic would likely spur additional inquiries, as has been the experience in other states. Developing a registry would provide a rich source of data, with which the collaborative could answer questions about care, service, and needs. The collaborative could begin to share the data they currently hold and identify pilot studies that might be parlayed into larger projects. The group would then be positioned to pursue grant funds for additional research. Hiring a team of research assistants would serve to jump start the research activities of senior researchers. After a couple of years, the research team should be grant-funded at a level that would allow the research team salaries to be covered by that external funding. The cost of funding a research assistant, a data manager, and a statistician to work half-time each would be $110,625.

Social Marketing Campaign

Any social marketing campaign for sickle cell disease should be coordinated with the objectives for the state plan. The campaign would be designed based on at least three factors: the message, target audience, and funding available for the campaign. When these factors are determined, then it will be possible to design a campaign to address the plan objectives.

There have been several marketing efforts in recent years that can form the foundation for a future campaign. The Department of Health has sent letters to parents of newborns whose babies test positive for sickle cell trait or disease. They currently send information about follow-up counseling at UAMS/Partners for Inclusive Communities. Partners has provided individualized education sessions with families of newborns, as well as more general public awareness through health fairs, meeting with community groups and churches, and outreach to health care providers. They have recently initiated an outreach campaign to the Hispanic community to make them aware that the condition exists within their families. Sickle Cell Support Services has run public service spots about sickle cell and has participated in a number of community events helping people to understand the condition. Arkansas Foundation for Medical Care has worked with physicians to help them learn how to best care for persons with sickle cell disease. In September 2010, the Arkansas Minority Health Commission will launch a major social marketing campaign to help people gain knowledge and understanding about sickle cell through television, radio, and print advertising with a toll-free number to call for more information.

Future efforts should be tailored to the audience and message. Options could include an additional general awareness campaign using television, radio, and newspaper advertising that builds on the campaign of the Commission; targeting physicians using direct mail and conferences for family practice physicians and pediatricians; and using schools to reach pre-teens for education about sickle cell trait and its implications for their lives.

The cost of creating advertising spots and purchasing media time and space would require $100,000 per year.

Recommendations

- Arkansas needs a Comprehensive Sickle Cell Program, in which children and adults can receive state of the art care and case management, regardless of where they live.

- The Program would utilize comprehensive, central centers ("Hubs") for both pediatric and adult patients, and multiple peripheral sites ("spokes") located throughout the state in areas where there are significant numbers of sickle cell patients.

- The Hubs would provide a comprehensive, multidisciplinary clinic; all patients involved would have at least one yearly comprehensive visit. Each visit would result in the development of a patient specific plan of care, which would be maintained by the center, the patient, and that patient's "Spoke" site. The comprehensive centers would be staffed by medical directors, and advanced practice nurses (APN) or nurse practitioner with specialized knowledge and/or experience in sickle cell disease, appropriate administrative and support staff; and would have available other services which are important in the care and management of sickle cell disease, such as pain management, social work, vocational training/support, etc. The Hubs will also develop a "24/7" call system, whereby patients, "Spoke" sites, or other medical professionals throughout the state can obtain immediate advice regarding the care of patients with sickle cell disease from trained personnel.

- The "Spokes" would be located in areas of the state with high concentrations of sickle cell patients, and would be available for interim management of acute complications of the disease, such as painful crises and infections. Spokes would be staffed by a willing physician provider (~0.1FTE) as well as a designated sickle cell nurse (~0.2 FTE), and would implement the patients specific plan of care, with appropriate backup from the Hub as needed.

- The state Medicaid office should be involved in the implementation of some or all of this plan, in that some revisions ("waivers") of the Medicaid program, specifically those limiting outpatient care and lab, and those limiting the number or prescriptions per month, are detrimental to the ability to deliver appropriate comprehensive preventive care to sickle cell patients. Additionally, aggressive, comprehensive case management of sickle cell patients, both children and adults, would result in not only improved care and quality of life for the patients, but also, based on the experience in other states, lead to significant potential for cost savings.

- A sickle cell disease educational outreach coordinator should be hired to help the agencies that provide educational services integrate their activities and to address unmet training needs.

- Creation of a sickle cell disease registry within the Arkansas Department of Health or the University of Arkansas for Medical Sciences.

- Mandatory reporting of sickle disease diagnosis and health status information to the registry by physicians, laboratories, hospitals, and the state Newborn Screening Program.
- The state should develop a research collaborative from agencies with an interest in examining outcomes for patients and families.
- Funding for a team of research assistants who would support principal investigators would provide a jump start for research studies. Under the supervision of senior researchers, the assistants could provide the time and expertise to collect, clean, and analyze existing data and provide the foundation for pursuing larger, grant-funded studies.
- Conduct a general awareness campaign using television, radio, and newspaper advertising.
- Target physicians using direct mail and conferences for family practice physicians and pediatricians.
- Use schools to reach pre-teens for education about sickle cell trait and its implications for their lives.

Potential Cost Savings

It is beyond the scope of this report to calculate strict cost utility estimates of the hospital and other costs that could come from reductions in either length of stay (an estimate is shown in Table 1), number of admissions, number of readmissions, or return to Emergency Department or hospital in 14 or 30 days. However, Table 1 shows an estimate of the savings to Arkansas Medicaid that might accrue if the lower charges found in non-metropolitan hospitals in 2008 were replicated in metropolitan hospitals. This artificial example is placed for illustration purposes only, to demonstrate the order of magnitude of possible cost savings.

			No.	Charges	Total est. charges
		Non-metro	167	8,924	$ 1,490,354.12
		Metro	719	13,871	$ 9,973,548.25
Target charges		Metro	719	8,924	$ 6,416,554.56
Savings opportunity					$ 3,556,993.69
Savings per discharge					$ 4,947.14
Est. savings to Medicaid					**$ 1,846,746.16**
Est. indigent care savings to Hospitals					$ 120,439.97

Table 1. Estimate of potential annual savings opportunity to Arkansas Medicaid from efficiencies in SCD hospital management.

Though this example is artificial, over the years, real savings have accrued to other hospitals and states that have instituted a case management system. For example, in Virginia, case management was implemented in 1993. Figure 1 shows that over the subsequent years, the number of patients returning to the ED for care within 30 days of a visit dramatically declined.

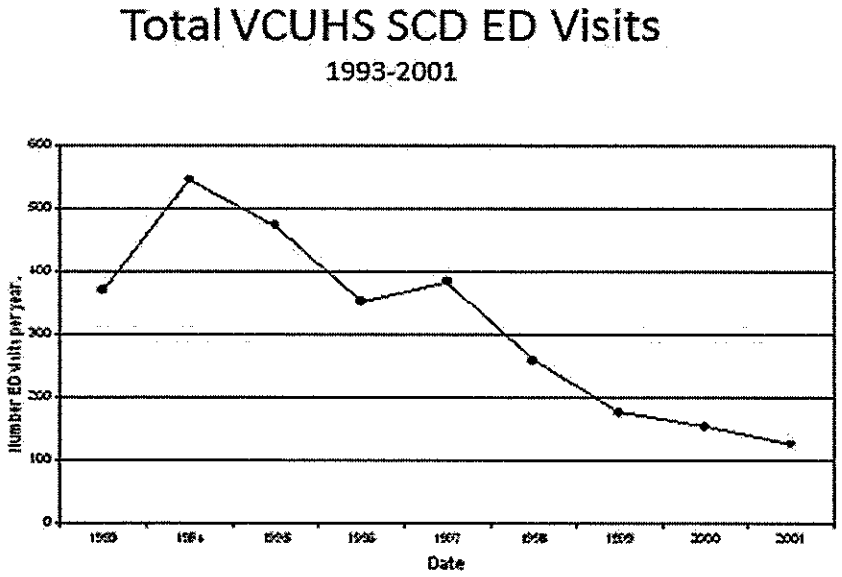


Figure 1. VCU Medical Center Sickle cell visits, seen in ED within 30 days of visit, 1993-2001

Figure 2 shows that, initially after case management began, the number of ED visits actually increased, perhaps because new patients were enrolled in the program in year 1. Subsequently however, for each year, the number of ED visits declined, sometimes dramatically from year to year. The overall reduction was approximately 50% over 6 years, and visit numbers were even lower in subsequent years. Figure 3 shows that this occurred while the number of patients enrolled in the program increased.

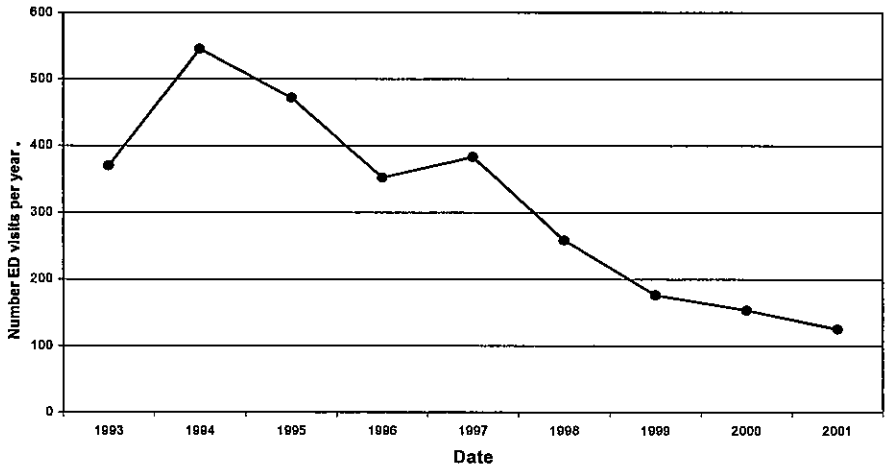


Figure 2. VCU Medical Center, number of sickle cell ED visits per year, 1993-2001.

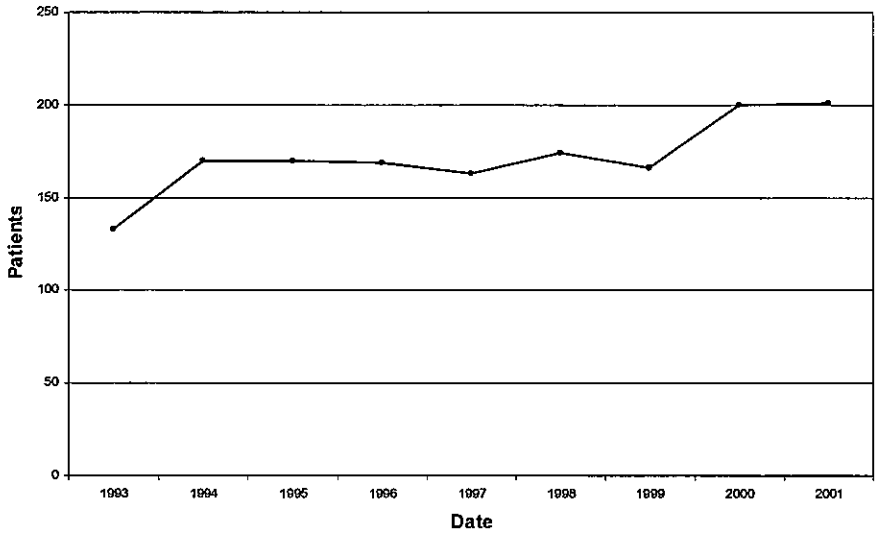


Figure 3. VCU Medical Center, number of sickle cell clients enrolled, 1993-2001.

In summary, through a case management and state consultation program instituted using a hub-and-spoke approach, Arkansas may have an opportunity to save up to $1.8 million annually in Medicaid costs (estimated) and/or reduce emergency department visits by nearly 50% (based on savings in other states) for sickle cell disease patients.

Attachment 1

Budget for System of Sickle Cell Clinics

Personnel	FTE	Cost/FTE	Salary for Project	Fringe	Salary plus fringe		Total Needed
Adult Hub							
Medical Director	0.5	$200,000	$ 100,000	$ 25,000	$125,000		
APN /NP	1.3	85,000	110,500	27,625	138,125		
Administration	0.5	50,000	25,000	6,250	31,250		
Social Worker	0.5	45,000	22,500	5,625	28,125		
Supportive Services						20,000	
Sub-total							**$342,500**
Pediatric Hub							
Medical Director	0.5	*No additional cost*					
APN /NP	1	*No additional cost*					
Administration	0.5	50,000	25,000	6,250	1,250		
Social Worker	0.5	45,000	22,500	5,625	28,125		
Supportive Services						20,000	
Sub-total							**79,375**
Spokes							
Nursing personnel	0.2	60,000	12,000	3,000	15,000		
Physician	0.1	200,000	20,000	5,000	25,000		
Social Worker	0.2	45,000	9,000	2,250	11,250		
Supportive Services						5,000	
Sub-total for one spoke						56,250	
Sub-total for four spokes							**225,000**
Training - 60-80 hours in 1st year							50,000
Total Cost - Year 1							**$696,875**

Budget for Sickle Cell Education Outreach

Personnel	FTE	Cost/FTE	Salary for Project	Fringe	Salary plus fringe	Total Needed
Outreach Coordinator	1	$65,000	$65,000	$16,250	$81,250	$81,250
Travel						5,000
Training Materials						5,500
Total						**$ 91,750**

Budget for Sickle Cell Registry

Category	Description	Cost	
Personnel	Software Support Analyst	$53,264	
	Health Program Specialist II	46,730	
	Fringe	30,351	
Out of State Travel	Conferences/Training	6,400	
In-state Travel		420	
Small equipment	Computers, workstations	8,000	
Office supplies	software, paper, etc.	4,500	
Large equipment	Server	15,000	
Other	Server maintenance contract	10,000	
Total			**$174,665**

Budget for Sickle Cell Research Infrastructure

Personnel	FTE	Cost/FTE	Salary for Project	Fringe	Salary plus fringe	Total Needed
Research Assistant	0.5	$55,000	$27,500	$ 6,875	$34,375	
Data Manager	0.5	42,000	21,000	5,250	26,250	
Statistician	0.5	80,000	40,000	10,000	50,000	
Total						**$110,625**

Budget for Social Marketing Campaign

Media Campaign-Purchase of Advertising	**$100,000**	

Total for All Activities **$1,173,915**