Expanding Comprehensive Services for Adults with Sickle Cell Disease

Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease

December 2008

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Report of the Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease: Executive Summary

Sickle Cell Disease (SCD) is a genetic blood disorder that affects more than 3,300 Marylanders. Over the past two decades, Maryland has made tremendous strides, through the use of comprehensive services, in *reducing mortality rates* and *improving the overall quality of life* for children and adolescents with SCD. Such comprehensive services, however, ***do not*** exist for many of Maryland's adults who are coping with SCD. The lack of comprehensive medical and social services for adults compromises their health and quality of life. Subsequently, it may lead to worse health outcomes and increased health care costs – the majority of which is financed by taxpayers. The Statewide Steering Committee on Services for Adults with Sickle Cell Disease was charged to address specific strategies and practices needed to facilitate the provision of comprehensive health care, help to improve the overall quality of life for adults with SCD, and reduce the public burden associated with health care costs for the uninsured and underinsured. In addressing the Committee's charge, issues surrounding access to care, reducing health care costs, health care services, and quality of life resources emerged.

The report is divided into five sections. The first provides an introduction to Sickle Cell Disease and the problem in providing services to adults with SCD. The second section reports on the committee charge, meetings, subcommittee formation, activities, and events. The third presents the issues surrounding the charge and reports findings of the committee's examination of these issues. The fourth section offers recommendations for continued improvement in services for adults with SCD. The final section concludes the report with recognition of the challenges involved and a request to continue its work as a Committee.

Summary of Committee Recommendations:

- Developing a statewide patient registry and identify a medical home for each patient
- Ensuring access to Medicaid or low cost private insurance
- Developing and promoting standard treatment protocols in emergency departments and other urgent care facilities
- Shifting resources toward comprehensive specialty care and preventive care models such as regional infusion centers
- Providing educational, employment, and psychosocial services to patients with SCD
- Increasing public awareness and education about sickle cell disease and sickle cell trait

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I. Introduction

What is Sickle Cell Disease? Why is it Relevant to Marylanders?

Sickle cell disease (SCD) is a complex inherited blood disorder in which structural changes in hemoglobin (a blood protein) causes red blood cells to become hardened, brittle, and sickle-shaped. These cells block the small blood vessels and obstruct the flow of oxygen and nutrients to various parts of the body – resulting in anemia, progressive organ damage, and severe pain. Sickle cell patients often experience a poorer quality of life, inhibited work productivity, and premature mortality compared to individuals without SCD.

There are approximately 1,600 children and adolescents with SCD in the state of Maryland. As a result of coordinated medical and social services that have been available over the past two decades, the mortality rate for young people in Maryland with SCD has *decreased* by a factor of ten. Consequently, Maryland's youth with SCD experience a developmental trajectory that is much healthier today compared to the late 1980's. More importantly, the advances in service provision also mean that more of Maryland's citizens are transitioning out of pediatric health care facilities and into adult care each year.

Unfortunately, medical and social services for adults with SCD in Maryland are not comparable to services and resources that are available to children. Current estimates indicate that more than 1,700 adults with SCD reside in the state of Maryland. Most adults do not receive or have access to the types of health care options that have been demonstrably effective in reducing morbidity and mortality among children and adolescents. This not only impacts the overall quality of life of adults with SCD, but the lack of comprehensive services also significantly contributes an additional tax burden on Maryland citizens. Public funds are often used to cover the health care expenses for adults with SCD who are either uninsured or insured by Maryland Medicaid. In October, 2007, The Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease was formed (Chapter 435, Acts of 2007) to explore ways for Maryland to address the service needs of adults with sickle cell disease.

The purpose of this report is to update the Maryland General Assembly on the progress and activities of the Committee.

II. Committee Charge and Deliberations

The Committee is charged with educating those with sickle cell disease (SCD), health care providers, and the public about State care and treatment. Also, the Committee is to establish partnerships with institutions and communities, as well as a statewide network of service providers for adults with SCD; a comprehensive education and treatment program for them; and a day infusion center. Moreover, the Committee will seek support for a campaign to educate health care providers regarding health disparities, community dynamics, cultural practices, behavioral and psychosocial issues, and standardized treatment and emergency room protocols.

The Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease is comprised of patient advocates, public health officials, health care providers, and academic researchers who are or work on behalf of persons coping with sickle cell disease

During its first year, the Committee met six times: November, 2007, January, February, March, May, and November, 2008. Co-chairs were decided upon: Dr. Donald Shell, Dr. Shawn Bediako, and Ms. Anika Wilkerson, who later resigned as co-chair but retained membership. Also, Sub-Committees were formed:

The Public Awareness Campaign Sub-Committee will explore promoting the use of standardized treatment guidelines, emergency room protocols and hydroxyurea monitoring protocols, and publicizing Medicaid's Employed Individuals with Disabilities Program to promote opportunities for health coverage;

The Community Based Support Group Activities Sub-Committee will look at establishing an effective SCD self-help support group;

The Sub-Specialist/PCP Education Awareness Campaign Sub-Committee will explore ensuring the availability of primary care by supporting primary care providers in their efforts to care for adult SCD patients, establishing an ongoing educational program for providers, establishing a 24/7 on-call consultant service, educating providers about the use of hydroxyurea, establishing a case management network to assist patients and support primary care physicians, and establishing a network of outreach and telemedicine clinics to complement the outreach case management network.

The Patient Registry Sub-committee will look at developing a web-based repository for an abbreviated electronic medical record, and using a confidential patient registry to ensure that all patients are receiving care consistent with established standardized guidelines. The sub-committee will contact and involve the several "registry projects" already underway at SCDAA, the NIH and MDLogix.

The Grant Sub-Committee will develop mechanism to obtain grants.

In addition, the Committee conducted or participated in several activities and events:

- The Sickle Cell Day with Legislators in Annapolis. The event was held on January 22 at the House Office Building, Room 302 from 9:00 a.m.-11:00 a.m. There was a good showing from legislators as well as community groups, families and friends. Attendees had the opportunity to meet with their legislators to discuss sickle cell disease and the continuum of care.

- Grand Opening of the Sickle Cell Infusion Center at Johns Hopkins on February 18, 2008. The center is located at 600 N. Wolfe Street, Carnegie Building.

- NIH Consensus Development Conference: Hydroxyurea Treatment for Sickle Cell Disease, February 25–27, 2008

- The National Transition Symposium

- Sickle Cell Disease Walk-a-thon on May 17, 2008 at 8:30 a.m. From Stanton Community Center, 92 W. Washington Street, Annapolis, MD 21401 to the Governor's Mansion.

- A local Maryland Division of Sickle Cell Disease of America has been established

III. Issues, Challenges, and Opportunities

In addressing the Committee's charge, issues surrounding access to care, reducing health care costs, health care services, and quality of life resources emerged. A discussion of Committee findings is presented in this section.

A. Expanding Services to Adults with Sickle Cell Disease

Access to Care. While Medicaid is the insurer for many adult SCD patients in Maryland, obstacles to enrollment persist. In order for an uninsured non-pregnant, adult SCD patient over the age of 21 and under the age of 65 without a dependant child to become eligible for Maryland Medicaid they must be declared a disabled person. Many adult SCD patients in Maryland are unable to meet the requirements for "disability" determination and therefore are unable to get access to Maryland Medicaid. Even though it is not necessary to qualify for SSI to obtain a favorable disability determination for Medicaid, the requirement for a disability determination by the Social Security Administration or by the State Review Team in the Department of Human Resources before admittance into Maryland Medicaid has and continues to serve as a protracted arduous task for many adults with SCD.

The Steering Committee recognizes that once Medicaid eligible; an adult sickle cell patient can receive access to all necessary medical services to prevent repeated uncompensated emergency department care and hospital admissions. Those who do not meet the eligibility requirements or are otherwise uninsured may use the system as uncompensated care patients. This system is inefficient, however, because Maryland continues to experience an increase in its financial obligation to cover costs associated with the delivery of uncompensated care. Uncompensated care, including bad debt and charity care, increased from $842 million in FY 2006 to $927 million in FY 2007[1]. The Steering Committee strongly recommends that the effort to contain costs, provide uninhibited access to quality hospital care, and meet standards of public accountability for persons coping with SCD must include the provision of Medicaid-covered preventive primary care and comprehensive specialty care to adults. This will prevent the more expensive and less effective frequent utilization of emergency department or inpatient hospital-based care.

An opportunity for expanded services exists in The Sickle Cell Treatment Act. On October 22, 2004, the Sickle Cell Treatment Act of 2003 was signed into law, amending title XIX of the Social Security Act and providing new optional benefits in the Medicaid program. It also makes matching Federal funds available for *education and outreach* to Medicaid-

eligible adults and children with SCD. The new provision offers several flexible options to states, including: (a) coverage of additional services that might not be covered in the state plan; (b) reimbursing SCD services at a different rate than that which is paid for similar services provided for other conditions; (c) establishing different coverage limits for SCD compared to those that apply to services in other benefit categories. Therefore, the Committee recommends the consideration of simplified qualification of adults with SCD for Maryland Medicaid, and the implementation and utilization of the provisions in the federal Sickle Cell Treatment Act affording additional resources and benefits to adults with SCD.

Reducing Health Care Costs. In 2005, Maryland's Medicaid program paid approximately $5,886 per member each month for the care of non-Medicare eligible Fee-For –Service adults with SCD. Hospital admissions accounted for almost 42% of this expenditure.[2] A significant reduction in these costs would be realized by investing in resources that keep SCD patients out of the hospital. Steering Committee members are in agreement that the optimal route to accomplish this goal is to ensure that individual health care needs of adults with SCD are addressed through regular preventive primary care and comprehensive specialty care. Adult SCD patients who are not eligible for Medicaid – but in need of health insurance – would benefit from subsidies that support their enrollment in plans similar to the Maryland Health Insurance Plans ("MHIP+") HMO product for limited or moderate income members. Premiums for this product range from $261 to $761 dollars per month for individual adults age 64 or below. Therefore, shifting emphasis towards services provided by specialty SCD clinics represent $5,100 to $5,600 in *savings* per member each month for Maryland's Medicaid program compared to costs for non-Medicaid eligible fee-for-service.

B. Health Care Services

Patient Registry and Patient Education. There is a need to develop and implement a statewide patient registry and personal electronic health record specifically for persons affected by SCD. The Committee proposes the development and implementation of a consumer-oriented Maryland Sickle Cell Disease Registry. This electronic case management registry should be developed based on health informatics standards and models such as provided by Health Level 7 (HL7), the Clinical Data Interchange Standards Consortium (CDISC), and the Public Health Data Standards Consortium (PHDSC). The personal electronic health record will not only utilize cutting-edge health information technology to facilitate continuity of care across health delivery systems, but will also house educational information and best practices-based case management tools that will assist clinicians, patients, and families in making informed decisions about the range of treatment options for SCD in ambulatory care settings. Use of these standards and models will enhance the clinical and public health capabilities of the system and will also strongly enable, with appropriate protections of patient privacy and confidentiality, use of the data for ongoing research to continue to improve knowledge and best practices.

Medical Home. The Committee recommends establishing a "medical home" model for adults with SCD where care is provided through coordinated collaboration between primary care and specialty physicians. Our medical home model includes the provision of evidence-based medical services that benefit patients, and if necessary, facilitates access to

additional psychological and/or social services. In this model, the physician is not limited to the role of managed-care gatekeeper; rather, she or he assists patients in obtaining appropriate services.

Standard Treatment Options. The Committee recommends enhanced outreach to educate physicians and health care professionals about the benefits of hydroxyurea, a chemotherapeutic agent shown to be an effective treatment for SCD pain. Recent data indicate that although clinical trials have established that consistent, appropriate use of hydroxyurea is associated with fewer hospital admissions, it is often underutilized.

Emergency Room Protocol. Currently, there is no standard uniform protocol for treatment of SCD in Maryland's emergency departments. Thus, some patients experience frustration, anger, and stigma when they seek emergency care during times in which their primary provider is not available. The Committee recommends the development of a standard treatment protocol – based on evidence-based best practices – that will ensure adherence to uniform procedures for treating acute SCD pain episodes no matter where in the state individuals with SCD present for care.

Day Infusion Center. The Committee recommends expanded support for the Sickle Cell Infusion Center at Johns Hopkins and the establishment of regional infusion centers across the state. The infusion center could help to reduce the number of hospital admissions. This would enable patients to more effectively manage their treatment, with minimal interference on activities of daily living. In addition, the net tax savings brought about by reduced expenditures associated with hospital admissions would outweigh the initial costs of supporting infusion centers. The Infusion Center at Johns Hopkins, which opened in February of 2008, is open during regular business hours, Monday through Friday. There have been well over 500 visits to the Center for infusions and an additional 200 urgent care visits. The admission rate for those patients seen for acute vaso-occlusive pain is 15%. This compares to the admission rate from the Johns Hopkins emergency department which is 33% during the week and increases to close to 60% on the weekends when the Infusion Center is closed. Providing additional resources to the Center for evening and weekend hours will further decrease hospital admission for sickle cell disease .

C. Quality of Life Resources

Employment and Educational Services. Current estimates indicate that adults with SCD are significantly less likely to have completed post-secondary education. Many adults with SCD experience difficulties in maintaining gainful employment. Given epidemiological studies that suggest relationships among education, employment, and health status, the Committee recommends the development of directed services that address these issues. Services include, but are not limited to, tutoring for GED preparation, occupational guidance counseling, job skills training, and education about patients' rights under the Americans with Disabilities Act.

Psychological Services. A well-established body of research reports that adults with SCD often experience elevated levels of stress, depression, and anxiety compared to the general population. These psychological factors could inhibit adherence to treatment

protocols and subsequently lead to a more advanced disease severity. Currently, there are very few mental health practitioners who are also knowledgeable about SCD. This significantly impacts the quality of psychological and counseling services that are offered to adult patients. The Committee recommends expanded outreach and education about SCD to licensed mental health providers who treat individuals with chronic conditions and the development of a statewide network of providers who are available to provide services to adults with SCD.

IV. Recommendations

The Maryland Statewide Steering Committee believes that significant improvements in the quality of life for adults living with SCD, along with significant savings for the State of Maryland may be achieved with the implementation of the recommendations included in this report. These recommendations are:

- Develop a statewide patient registry to facilitate continuity of care across health care systems and providers
- Ensure the provision of Medicaid or low cost private insurance for both preventive primary care and comprehensive specialty care to adults with SCD
- Educate physicians and other health care professionals, including mental health providers, on best practices in the management and treatment of SCD
- Shifting resources toward comprehensive specialty care and preventive care models such as regional infusion centers
- Use the range of services provided by the Sickle Cell Infusion Center at Johns Hopkins (i.e., continuity of care and access to psychiatric/social services) as a model to be implemented in other regions of the state
- Develop a standard protocol for SCD treatment in emergency department and other urgent care settings
- Enhance patient education to include appropriate pain self-management and information about the range of evidence-based treatment options
- Increase the availability of educational, employment, and psychosocial counseling services for adults with SCD
- Increase public awareness and education about sickle cell disease and sickle cell trait

V. Conclusion

Maryland has made significant progress in the care and treatment of its pediatric sickle cell disease patients. However, there are challenges to be met in offering similar services to adults. The activities of the Maryland Statewide Committee on Services for Adults with Sickle Cell offer the first step toward improving the health outcomes and overall quality of life of the more than 1,700 adults in Maryland who are coping with SCD. Specifically, the Committee has included membership representatives from all groups as outlined in the law; institutional and community partnerships, and a network of stakeholders have been established; efforts to educate the public, sickle cell disease patients, and their providers about the care and treatment of adults with Sickle Cell Disease have been initiated; a subcommittee to seek grant funding has been formed; development of recommended standard and emergency treatment protocol has begun; and a day infusion center has been established at Johns Hopkins, opening in February, 2008. The Committee hopes to continue the public dialogue and work needed to advance these improvements presented in this report.

Maryland Statewide Steering Committee on Services for Adults

with Sickle Cell Disease

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[1] Health Services Cost Review Commission (HSCRC). "Average Amount Paid For A Hospital Stay in Maryland". September 10, 2008.

[2] Department of Health and Mental Hygiene 2006 Legislative Report: The Study of Adult Sickle Cell Disease in Maryland
http://www.fha.state.md.us/pdf/genetics/2006sicklecell_legis_rpt24.pdf