

## Short Communication

# Improving the Healthcare Model for Management of Adults with Sickle Cell Disease in the PPACA Era

Julie Kanter<sup>1\*</sup> and Lanetta B. Jordan<sup>2</sup><sup>1</sup>Department of Pediatric Hematology-Oncology, Medical University of South Carolina, USA<sup>2</sup>Department of Public Health Sciences, University of Miami, USA

## \*Corresponding author

Julie Kanter, Medical University of South Carolina, Charleston, SC 29483, Tel: 1-843-876-8483; Fax: 1-843-792-8912; Email: kanter@musc.edu

Submitted: 08 December 2014

Accepted: 18 July 2015

Published: 27 July 2015

ISSN: 2333-6684

## Copyright

© 2014 Kanter et al.

## OPEN ACCESS

## Keywords

- Pediatric
- Adult
- Health outcomes
- Insurance
- Sickle cell disease

## Abstract

Sickle cell disease (SCD) is the most common inherited blood disorder and affects approximately 100,000 individuals in the United States. Once confined to childhood, improvements in early diagnosis through universal newborn screening and coordinated pediatric programs for SCD have contributed to an increased lifespan, with patients surviving into adulthood. However, there is a paucity of healthcare resources available for the adult patient population.

SCD has become a chronic condition as patients live into adulthood; therefore, the paradigm of care needs to shift from managing acute conditions to preventing complications and preserving health. Unfortunately, the limited numbers of knowledgeable and available providers, as well as a disorganized healthcare infrastructure, have caused most affected adults to rely on acute care services, leading to increased rates of hospital utilization and higher healthcare costs. For adult patients with SCD (and other chronic conditions) who lack third-party payer coverage, the Patient Protection and Affordable Care Act (PPACA) may alleviate some of these issues in states accepting Medicaid expansion. However, while the PPACA may provide for increased insurance coverage for some patients, it will not increase the number of available providers or improve health outcomes for this patient population. Thus, it is necessary to create a structured system of care in the United States to manage patients with SCD and their long-term comorbidities and complications.

**Summary:** A healthcare model that provides a coordinated system of healthcare delivery for adults with SCD is the first step to improve health outcomes.

## ABBREVIATIONS

SCD: Sickle Cell Disease; HTC: Hemophilia Treatment Center; PPACA: Patient Protection and Affordable Care Act

## INTRODUCTION

Sickle cell disease (SCD) is the most common inherited red blood cell disorder affecting individuals of African, Mediterranean, and Asian descent. SCD is caused by a single point mutation on the  $\beta$ -globin subunit of hemoglobin, resulting in fragile, sickle-shaped red blood cells with a reduced lifespan [1]. The most common SCD genotypes in the United States are hemoglobin SS and SC, and sickle  $\beta$ -thalassemia ( $S\beta^0$ ,  $S\beta^+$ ) [1]. However, even among patients with the same genotype, SCD shows considerable phenotypic heterogeneity. Patients experience a range of symptoms and complications, including acute pain (vaso-occlusive crisis), chronic pain, infections, stroke, acute chest

syndrome, and end-organ damage, due to chronic hemolysis, microinfarction, and vaso-occlusion [1,2]. With the advent of universal newborn screening, early treatment of infection, and newer immunizations, the life expectancy of patients with SCD has markedly increased such that SCD is no longer confined to childhood and has become a chronic disease in adults [2]. This is evidenced by a significant improvement in life span between 1979 and 2006 for patients with SCD [3].

As more patients are living into adulthood, management of SCD must shift from the paradigm of acute management of complications to a chronic care model that focuses on prevention of common complications, as well as on early identification of and intervention for these complications. In this communication, we examine the challenges imposed on adult patients with SCD by the current health system in the United States and offer an

alternative coordinated care model for effective healthcare delivery in this patient population.

### Disease Complications

As patients with SCD age, they face an increased number of disease-related comorbidities and complications (eg, vaso-occlusive pain, hemorrhagic stroke, and multi-organ damage) [2,4]. They often develop more chronic pain, compared with the intermittent pain of childhood, which requires more intense treatment and specialization. Patients also develop multi-organ dysfunction and require evaluation and treatment by providers from multiple specialties, which can necessitate significant time and care coordination [2].

### Access to Appropriate Care for Patients with SCD in Adulthood

Although comprehensive care programs exist for pediatric patients with SCD, there is a paucity of accessible, trained providers for adults [2]. Also, patients with SCD often have publicly funded insurance (ie, Medicaid or Medicare) [5], and the third-party payer may be a barrier for patients because many adult healthcare providers limit the number of publicly insured patients they can accept. Simultaneously, as a patient with SCD transitions from adolescent to adult care, the system of care becomes less deliberate and more fragmented [2].

Lack of accessible healthcare providers may be a contributing factor to the inappropriate utilization of emergency department services, higher inpatient hospitalizations, and shorter time to rehospitalization in adult patients (Figure 1) [4,6]. The Healthcare Cost and Utilization Project, for example, showed that more hospitalizations for SCD occurred among patients aged 18 to 44 years (66%) than among those aged 1 to 17 years (24%) [5]. Similar results were reported in another study in which the highest acute care utilization, including rehospitalization and emergency department return visits, occurred in patients aged 18 to 30 years [4].

### Healthcare Utilization as a Measurement of Coordinated Care

Increased utilization of emergency department services and hospitalizations may be a marker of poorer care in adults with SCD. A recent study of Medicaid data from 5 states demonstrated that adult patients with SCD had greater emergency department reliance, more inpatient hospitalizations, and higher overall healthcare costs compared with pediatric patients with SCD [5]. Although not proven in this patient population, it is likely that the increased reliance on emergency services is a result of lack of access to appropriate primary care or disease-specific care centers [4]. Primary care physicians are often uncomfortable managing chronic pain and complications of SCD without the guidance of SCD specialists [7]. Lack of co-management for SCD may result in unnecessary blood transfusions (eg, for baseline anemia) [8], and complications may be mistaken for acute vaso-occlusive episodes, which can delay diagnosis and treatment of underlying complications [8]. Studies in other adult chronic diseases (hypertension, diabetes mellitus, and heart failure) have demonstrated that a deliberate approach to preventive, acute, and follow-up care resulted in more appropriate utilization of

healthcare resources and an improved quality of life for patients, suggesting the importance of quality coordinated care [9].

### Appropriate Model of Care

Healthcare models used in cystic fibrosis and hemophilia (conditions that require lifelong multidisciplinary care) may provide helpful frameworks for SCD. For example, hemophilia treatment centers (HTCs) employ a comprehensive care model by offering medical treatment with physician and nursing specialists as well as social work, case management, and physical therapy. There are approximately 141 HTCs [10] and programs throughout the United States for 20,000 patients with hemophilia [11]. HTCs treat individuals with this bleeding disorder throughout their lives via continuous supervision of all medical and psychosocial aspects of care [10]; according to one estimate, 67% of patients with hemophilia are treated at HTCs [12]. Patients with hemophilia who are treated at HTCs are 30% less likely to die of hemophilia-related complications compared with those not managed at HTCs [12].

Treatment centers like these have not been developed for SCD. The previously funded National Institutes of Health, National Heart, Lung, and Blood Institute Comprehensive Sickle Cell Centers were predominantly pediatric centers and focused on research, not care delivery [13]. The Health Resources and Services Administration SCD demonstration projects have not provided consistent, sustainable funding to grantee sites and offer only limited, highly competitive awards. Current SCD centers do not have sufficient funding to offer the coordinator resources of an HTC. Instead, these clinics often make use of community-based coordinators or social workers, community educators, and outside therapists instead of more structured, disease-directed care. Additionally, while centers for cystic fibrosis and hemophilia often have means for treating both pediatric and adult patients, those for SCD are often not well synchronized, which limits the efficacy of lifespan care. Dedicated, disease-specific care that includes physicians, case management, social support resources, and nurse education for patients with SCD would greatly enhance both quality care and the ability to obtain prospective data to better inform disease management and prevention of complications.

### Patient Protection and Affordable Care Act

There are several positive features of the Patient Protection and Affordable Care Act (PPACA) that will affect the care of patients with SCD. The new policy will remove the \$40,000 claims cap so that insurance companies cannot limit the cost of lifetime treatment [14], and the PPACA will eliminate the issue of preexisting conditions in the attainment of health insurance [14]. Finally, in states expanding Medicaid, the PPACA may extend coverage to additional adults with SCD who qualify [14]. However, many of the early implementation aspects of the PPACA, including the lack of several states' acceptance, will slow the positive enrollment projections of legislation for patients with SCD. Additionally, the PPACA does not alter the current reimbursement for certain medical services, such as blood transfusions, education, and counseling or case coordination; therefore, it will not have an impact on the ability to bill for services currently being rendered by physicians who care for

patients with Medicaid. The PPACA will not increase access to care for patients with SCD because it will not increase the number of physicians willing and able to treat affected patients. Because the PPACA will not universally increase reimbursement for care coordination, physicians will continue to be challenged in caring for complicated patients, such as those with SCD. Thus, alternative methods for improving care for patients with SCD should be explored and implemented. These mechanisms could include value-based incentives for physician practices willing to care for persons with SCD with demonstrated improved outcomes, bundled care payment systems for SCD centers, and/or enhanced disease-specific opportunities for education to deliver quality care.

## SUMMARY

SCD is a chronic condition that requires access to knowledgeable practitioners and a coordinated system of care throughout the patient's lifespan [2]. The system of care and payment system for adult patients with SCD should change to support the chronic care model. The PPACA will not enhance access to care for individuals with chronic conditions; it will only expand coverage to a percentage of patients [14]. Patients with SCD are living longer, but the system of care must change to ensure they are living better. Currently, it is difficult to obtain data on the care and outcomes of adult patients with SCD owing to the significant fragmentation of the care system [3,6]. Models of care that are focused around treatment centers, such as those effectively used in hemophilia and cystic fibrosis [9], are more likely to bring about true improvements in quality care as well as lay the groundwork for assessing the effect on actual patient outcomes.

## ACKNOWLEDGEMENTS

Under the direction of the authors, Susan M. Cheer, PhD, and Susan M. Kaup, PhD, of Evidence Scientific Solutions provided editorial assistance.

## Conflict of Interest

Julie Kanter, MD, received a grant from Hyundai Hope on Wheels and payment for lecturing from the American Academy of Pediatrics. Lanetta B. Jordan, MD, MPH, MSPH, received consulting fees as a speaker for Novartis Pharmaceuticals Corporation.

## REFERENCES

1. Chakravorty S, Williams TN. Sickle cell disease: a neglected chronic disease of increasing global health importance. *Arch Dis Child*. 2015; 100: 48-53.
2. Blinder MA, Vekeman F, Sasane M, Trahey A, Paley C, Duh MS. Age-related treatment patterns in sickle cell disease patients and the associated sickle cell complications and healthcare costs. *Pediatr Blood Cancer*. 2013; 60: 828-835.
3. Hassell KL. Population estimates of sickle cell disease in the U.S. *Am J Prev Med*. 2010; 38: S512-521.
4. Brousseau DC, Owens PL, Mosso AL, Panepinto JA, Steiner CA. Acute care utilization and rehospitalizations for sickle cell disease. *JAMA*. 2010; 303: 1288-1294.
5. Steiner CA, Miller JL. Sickle cell disease patients in U.S. hospitals. 2004. HCUP Statistical Brief #21. December 2006. Agency for Healthcare Research and Quality, Rockville, MD. <http://www.hcup-us.ahrq.gov/reports/statbriefs/sb21.pdf>.
6. Kauf TL, Coates TD, Huazhi L, Mody-Patel N, Hartzema AG. The cost of health care for children and adults with sickle cell disease. *Am J Hematol*. 2009; 84: 323-327.
7. Crosby LE, Quinn CT, Kalinyak KA. A biopsychosocial model for the management of patients with sickle-cell disease transitioning to adult medical care. *Adv Ther*. 2015; 32: 293-305.
8. Bender MA, Douthitt Seibel G. Sickle cell disease. In: Pagon RA et al, eds. *GeneReviews*®. Seattle (WA): University of Washington, Seattle; 1993-2015.
9. Wagner EH, Austin BT, Davis C, Hindmarsh M, Schaefer J, Bonomi A. Improving chronic illness care: translating evidence into action. *Health Aff (Millwood)*. 2001; 20: 64-78.
10. National Hemophilia Foundation. *Comprehensive Medical Care: Hemophilia Treatment Centers*. 2014.
11. Zhou ZY, Riske B, Forsberg AD, Ullman M, Baker JR, Koerper MA, et al. Self-reported barriers to hemophilia care in people with factor VIII deficiency. *Am J Prev Med*. 2011; 41: S346-353.
12. Soucie JM, Nuss R, Evatt B, Abdelhak A, Cowan L, Hill H, et al. Mortality among males with hemophilia: relations with source of medical care. The Hemophilia Surveillance System Project Investigators. *Blood*. 2000; 96: 437-442.
13. Sickle Cell Information Center. 2015.
14. U.S. Department of Health & Human Services Key features of the Affordable Care Act by year. 2014.

### Cite this article

Kanter J, Jordan LB (2015) Improving the Healthcare Model for Management of Adults with Sickle Cell Disease in the PPACA Era. *J Hematol Transfus* 3(1): 1037.