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Abstract

This paper states the need to declare Sickle Cell Disease as a top-level health priority. It calls for Sickle Cell to cease being a victim of institutional and societal discrimination. It provides an expansive overview of the disease and how society continues to treat it with inequity relative to other genetic disorders like hemophilia and cystic fibrosis, conditions that affect far fewer people. The paper also reveals and discusses a prevailing pattern of unequal treatment that is evidenced by disparities in research, government support, health care, health care insurance, social service support and advocacy. Finally, this paper presents recommendations that, if implemented, would begin to rectify the current disparate status of the world's most common genetic disease.

SICKLE CELL DISEASE: THE ULTIMATE HEALTH DISPARITY

Introduction

The topic of health disparities has become commonplace in the health care, social service and political fields. By its very nature, this topic is confusing on a number of levels and finding consensus on a definition is difficult at best. According to the National Conference of State Legislatures, "The term 'health disparities' refers to population-specific differences in the presence of disease, health outcomes, quality of health care and access to health care services that exist across racial and ethnic groups. Disparities represent a lack of efficiency within the health care system and therefore account for unnecessary costs."¹

Dr. Darrel G. Kirsh, President of the Association of Medical Colleges, clarifies the importance of addressing health disparities in his letter on the association's website. Dr. Kirsh explains, "Health disparities directly reflect social determinants of health and are higher in

communities where there is greater poverty, more stress, less access to fresh foods, and less recreational space. Disparities are also connected to increased medical errors, prolonged hospital stays, avoidable hospitalizations and readmissions, over and under-utilization of procedures, and failures in timely referral to specialist care. Given these factors, reducing health disparities is more than just the right thing to do—it is a critical component of improving health care quality.”²

In addition to improving health care quality, reducing health disparities can lead to a reduction in health care costs. This is true because many of the people who are subjected to health disparities end up utilizing more intensive and more costly emergency room and in-patient hospital services as a result of their inability to easily access preventive health maintenance

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Dr. Darrel G. Kirsh

services at the same levels and rates as those who do not suffer from disparate health care. Of particular importance in this regard are those who suffer from Sickle Cell Disease (“SCD”).

An article posted by Teresa L. Kauf in 2009 in the American Journal of Hematology reported that the annual cost of medical care in the US for people who suffer from SCD exceeds \$1.1 billion.³ Obviously, the eradication of SCD would lead to the elimination of this tremendous economic burden on our nation’s economy. However, reductions in the health disparities involved in SCD would also lead to significant reductions in SCD health care costs, most of which are borne by the federal

government through the Medicaid and Medicare programs.

Federal laws prohibit discrimination on the basis of race, religion, color, sex (including pregnancy and gender identity), national origin, age, disability, family medical history, or genetic information. These protections are typically applied to employment and housing. It seems only natural, then, that discrimination that exists in the form of the preferential treatment of one genetically transmitted disease over another should also be prohibited.

It is the intent of this paper to encourage the elevation of SCD to a higher level of importance in the health disparities discussion in order to attract the attention and resources that other health conditions enjoy as “official” health disparity topics. In order to do so, we must first examine the issue of why SCD is not officially recognized as a health disparity topic by the Centers for Disease Control and Prevention (“CDC”). We will also need to review the different aspects of inequity impacting SCD patients, including disparities in research, government support, medical care, medical insurance, social services and advocacy.

About Sickle Cell Disease

SCD is one of the most common genetically transmitted diseases on Earth. Estimates indicate that there are as many as 2,000,000 people worldwide and approximately 100,000 in the United

States who suffer from SCD. It occurs most frequently in people who live in or have descended from parts of the Earth's tropical and sub-tropical regions where malaria is or has been present. It is carried mainly by people with origins in Africa, South and Central America, the Caribbean, and the Middle East. Although SCD is most commonly found in people of color, it also affects a small percentage of Caucasians.

SCD is caused by an abnormal type of hemoglobin called hemoglobin S. Hemoglobin is a protein inside red blood cells that carries oxygen. Red blood cells carry oxygen to the body and are normally shaped like a disc. SCD is a genetic disorder in which hemoglobin S causes red blood cells to form an abnormal sickle or crescent shape. The fragile, sickle-shaped cells deliver less oxygen to the body's tissues and they create blockages in the patient's blood vessels. These problems decrease the amount of oxygen flowing to all parts of the body beyond the blockage and, over time, lead to severe and irreversible damage to tissue, organs bones and joints.

Wikipedia, the open-source online encyclopedia, gives a very accurate overview of what happens when the patient's red blood cells break down during what is known as a "Sickle Cell Crisis." It states, "The vaso-occlusive crisis is caused by sickle-shaped red blood cells that obstruct capillaries and restrict blood flow to an organ, resulting in ischaemia, pain, necrosis and often organ damage. The frequency, severity, and duration of these crises vary considerably. Painful crises are treated with hydration, analgesics, and blood transfusion; pain management requires opioid administration at regular intervals until the crisis has settled. For milder crises, a subgroup of patients manage on NSAIDs (such as diclofenac or naproxen). For more severe crises, most patients require inpatient management for intravenous opioids; patient-controlled analgesia (PCA) devices are commonly used in this setting. Vaso-occlusive crisis involving organs such as the penis or lungs are considered an emergency and treated with red-blood cell transfusions"⁴

With its disruptive occurrences of severe pain, frequent hospitalizations and the overabundance of medical complications that it produces, SCD can make it a monumental challenge for its sufferers to do things that most people take for granted.

There are many clinical manifestations of SCD and they vary dramatically in prevalence, frequency and severity between individual patients. Among the complications are: frequent infections, stroke, pneumonia, acute chest syndrome and thoracic cage infarction, osteomyelitis, avascular necrosis, heart disease, chronic renal failure, septicemia, meningitis, retinopathy, gallstones, hepatomegaly and chronic leg ulcers.

SCD greatly impacts the lives of those who suffer from it. It also affects their families, friends and neighbors. With its disruptive occurrences of severe pain, frequent hospitalizations and the overabundance of medical complications that it produces, SCD can make it a monumental challenge for its sufferers to do things that most people take for granted. SCD makes it more difficult to complete schooling, obtain and maintain employment, participate in and enjoy social

functions and even establish and nurture social relationships. SCD can be, in effect, a prolific “game changer” that is constantly stacking the deck against its carriers in multiple ways.

SCD Not Recognized as CDC Health Disparity Topic

In its 2011 “Health Disparities and Inequalities Report,” the CDC states, “In recent decades, the nation has made substantial progress in improving U.S. residents’ health and reducing health disparities, yet health disparities by race and ethnicity, income and education, disability status, and other social characteristics still exist.”⁵ In its own words, this landmark report “addresses disparities in health care access, exposure to environmental hazards, mortality, morbidity, behavioral risk factors, disability status and social determinants of selected health problems at the national level.”⁶

Due credit should be provided to the CDC for the effort to call out these disparities yet its list does not include SCD, a condition that arguably represents the ultimate of all health disparities.

The CDC’s report included twenty-two (22) health disparity topics that were chosen because they met one or more criteria. These criteria included, “Leading cause of premature death among certain segments of the U.S. population; social, demographic, and other disparities in health outcomes exist; effective and feasible interventions exist to address health outcomes; and high quality data were readily available from national health monitoring systems.”⁷ Due credit should be provided to the CDC for the effort to call out these disparities yet its list does not include SCD, a condition that arguably represents the ultimate of all health disparities.

The CDC’s report includes analytic essays grouped by six (6) different categories. Those categories are; 1) Social Determinants of Health; 2) Environmental Hazards; 3) Health-Care Access and Preventive Health Services; 4) Health Outcomes – Morbidity; 5) Health Outcomes – Mortality; and, 6) Health Outcomes – Behavioral and Risk Factor. Some of the topics included in the “Health Outcome – Morbidity” category are Obesity, Pre-term Births, Potentially Preventable Hospitalizations, Current Asthma Prevalence, Diabetes, Asthma, HIV/AIDS and Hypertension.

The absence of SCD from the CDC’s list of disparities is worthy of discussion, especially since it is listed as “Priority 4” in the CDC’s National Center on Birth Defects and Developmental Disabilities strategic plan.⁸ The CDC identifies Priority 4 as “Preventing and controlling complications from hemoglobinopathies, like Sickle Cell Disease (SCD) and Thalassemia.” This demonstrates that the CDC has established some level of commitment to SCD. However, it has elected not to elevate it to its overall list of health disparity topics.

SCD and its complications drastically reduce life expectancies when compared to the general population. There is a tremendous lack of medical knowledge about SCD and inadequate levels of experience and preparedness for treating it within the health care industry. In addition, the lack of medical provider readiness has a direct and profound effect on health outcomes.

Therefore, it appears that SCD should qualify as a health disparity based on the criteria utilized by the CDC.

An American Academy of Pain Medicine paper published in 2003 by Dr. Carmen R. Green and others says that pain management protocol for SCD patients is clearly identified as disparate. “Chronic, interepisodic pain is treated on an outpatient basis with analgesic medications and is often incompletely controlled, with patients and medical care providers alike expressing concern over patients’ long-term reliance on opioid analgesic medications. Pain remains the leading cause for ER visits and hospitalizations for people with SCD. The above findings, suggesting disparities in analgesic administration, raise concerns regarding the care that patients experiencing severe acute SCD pain may encounter.”⁹

Dr. Green and her colleagues are not alone in their assertions of health care disparities related to SCD. A paper published in 2005 by Doctors Lauren A. Smith, et al, entitled, “Sickle Cell Disease: A Question of Equity and Quality,” describes another specific example of the disparate health care treatment SCD patients are confronted with. Dr. Smith and her co-authors state, “The limited evidence on the quality of SCD care suggests that the significant gains in clinical care for SCD have not been uniformly distributed. For example, although penicillin prophylaxis is effective in preventing morbidity and mortality because of invasive pneumococcal disease, recent data demonstrate inadequate prophylaxis rates among publicly insured children compared with their privately insured counterparts.”¹⁰

“The limited evidence on the quality of SCD care suggests that the significant gains in clinical care for SCD have not been uniformly distributed.”

Dr. Lauren Smith, et al.

Additionally, Smith’s paper declares, “The question of race has been inextricably linked with SCD since its recognition as a distinct disease. Although it is uncomfortable to contemplate, we must consider the possibility that conscious or unconscious racial bias adversely affects the availability of resources not only for research and the delivery of care, but also for the improvement of that care.”¹¹ Dr. Smith, et al, also assert, “It is tragic and unjust for a particular group of patients to suffer avoidable complications and even death because effective new therapies have not been uniformly implemented. Although this is not unique to SCD, the severity of the disease and the nature of who suffers from it make the impact of this failure both severe and disparate.”¹² If health outcomes are among the criteria for inclusion in the CDC’s list of health disparities, it would seem that SCD should qualify based on the information cited in the above paragraphs.

Another of the criteria for inclusion on the list was the “existence of effective and feasible interventions to address health outcomes.” In view of the significant increases in life expectancy that have occurred over the last two decades, there is no doubt that there have been drastic improvements in the ability to treat SCD patients. Thus, it is obvious that there is at least some level of treatment effectiveness currently in place, although, as Smith states, this level of

effectiveness is not necessarily common across the entire spectrum. Yet, all things considered, it appears that SCD clearly meets the criteria of having effective interventions in place to address health outcomes.

The final criteria for inclusion on the CDC list of health disparities was for high quality data to be readily available from national health monitoring systems. There is little doubt that SCD could not qualify for this particular criteria. A simple Internet search will show that, among other things, there is no agreement on the actual number of SCD patients residing with the United States. Estimates typically range from 80,000 to 100,000 people. Some estimates are lower and some are even as high as 130,000. For years, organizations like the Sickle Cell Disease Association of America and others have called for a national SCD registry in order to more clearly establish the number of SCD sufferers.

“In comparison, cystic fibrosis has enjoyed greater governmental and philanthropic support despite Sickle Cell Disease being 2.5 times more common. Cystic fibrosis is a genetic disease of whites.”

Ahmad Hazam, et al.

In the April 2011 “Newsletter of the International Society for Evidence-Based Health Care,” Authors Ahmad Hazam, Victor Montori and M. Hasan Murad wrote, “The National Heart, Lung and Blood institute is in the process of developing clinical practice guidelines to improve the care of patients with Sickle Cell Disease. In support of this initiative, methodologists from the Grading of Recommendations Assessment, Development and Evaluation (GRADE) Working Group are assisting in conducting over 20 systematic reviews to summarize the evidence and help facilitate the development of evidence-based guidelines. This process has uncovered a disheartening disparity. Despite the societal and personal burden, and the fact that 2011 marks the centennial anniversary of describing the phenotype and clinical presentation of the disease, the quality and quantity of evidence in this field remains minimal compared

to diseases of similar burden and prevalence in Caucasians or high income countries.”¹³ The authors also affirm, “In comparison, cystic fibrosis has enjoyed greater governmental and philanthropic support despite Sickle Cell Disease being 2.5 times more common. Cystic fibrosis is a genetic disease of whites.”¹⁴

A May 2003 paper prepared by the University of Rochester’s Kevin Fiscella entitled, “Assessing Health Care Quality for Minority and Other Disparity Populations,” provides additional confirmation about the lack of SCD related high quality evidence. In the paper, Fiscella reports, “Some of the conditions relevant to minorities that are not adequately addressed by existing measures have been previously identified. These include asthma, maternal/child care, pain management, HIV/AIDS, low back pain, sickle cell anemia, mental health, end-of-life issues, and cultural competency.”¹⁵ Fiscella also maintains, “Minority children with special health care needs are less likely than whites to have seen a physician but more likely to be hospitalized during the past year. Specific quality indicators are needed to monitor the care provided to these children with special needs. Examples of these conditions include congenital heart disease,

cerebral palsy, chronic renal failure, sickle cell anemia, cystic fibrosis, severe asthma, childhood cancer, major mental illness, severe developmental delay, and mental retardation.”¹⁶

It would therefore seem that SCD meets at least three of the four criteria established by the CDC for inclusion on its list of health disparities. First, there is no question that the average life expectancy of SCD patients is estimated to be approximately 30 years shorter than the national average. As such, it would appear that SCD should meet the criteria for a “leading cause of premature death among certain segments of the U.S. population.” Second, it appears that “social, demographic, and other disparities in health outcomes exist” in regards to SCD as a result of non-standardized treatment protocols practiced around the country. Third, advances in early detection, penicillin and hydroxyurea therapies have been credited with extending the life expectancies of SCD patients. This fact should qualify SCD for the “effective and feasible interventions exist to address health outcomes” criteria. By these three measures, it would seem that SCD belongs on the health disparities list. The question remains, “why has SCD been excluded?” Those in the SCD field should be asking this question of the CDC and Congress until a suitable answer is received or until the list of disparities is amended to include SCD.

Disparity in Research

A review of the history of SCD research shows a sustained pattern of disparity of alarming proportions. Since its identification in 1910 until the passage of the National Sickle Cell Control Act in 1972, approximately \$1 million had been spent on SCD research. That equates to a little more than \$16,000 per year. The situation drastically improved with the passage of that legislation yet SCD research is still not on par with other diseases that affect far fewer people. Dr. Lauren Smith notes that a glaring disparity exists between SCD and Cystic Fibrosis research.

She claims, “Although per capita expenditures do not fully capture the differing experiences of disease by individuals, it is notable that NIH [National Institutes of Health] allocates almost 4 times more funding per person affected with cystic fibrosis as it does for those affected by SCD. These levels of funding have been essentially stable over the past 4 years.”¹⁷ The true extent of this disparity is found in the fact that SCD affects approximately 100,000 individuals and Cystic Fibrosis affects only about 30,000 persons.

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Dr. Lauren Smith, et al.

Cystic Fibrosis is not the only disorder that eclipses research funding for SCD. Research expenditures for Alzheimer’s Disease will be \$498 million; Asthma research will total \$221 million; Cystic Fibrosis will reach \$79 million; Hepatitis-C research spending will be around \$113 million; and Lupus research will top out over \$105 million; Multiple Sclerosis will be \$121 million; and Parkinson’s Disease research will reach \$151 million. In comparison, the money spent on research in 2012 for SCD is expected to amount to \$65 million.¹⁸

The National Human Genome Research Institute (“NHGRI”) in Bethesda, Maryland conducts research on genetically transmitted diseases. Its website lists forty-eight (48) genetic, orphan and rare diseases. NHGRI is conducting research on 19 of them.¹⁹ Among the disorders being researched are Cystic Fibrosis, Charcot-Marie-Tooth Syndrome, Gaucher Disease and Neurofibromatosis. Although it is listed as one of the qualifying disorders, NHGRI is not currently engaged in SCD research. In that SCD is widely accepted as the most predominant genetically transmitted disease in the world and in that NHGRI’s mission states that it “supports the development of resources and technology that will accelerate genome research and its application to human health,” it is peculiar that it is not actively involved in SCD research. NHGRI may certainly have good reasons why it is not pursuing SCD research but no such explanations are offered on its website.

“SCD is considered an ‘orphan disease’ (poorly publicized and not yet adopted by the medical research and funding communities) because it affects fewer than 200,000 people nationwide. Although the medical burden of orphan diseases is great, these diseases often are last to receive research funding.”

National Heart Lung and Blood
Institute Report

It is said that the fact that SCD is an “orphan disease” is one of the primary reasons that research on it lags behind that of other diseases. That is what is stated in the National Heart Lung and Blood Institute’s 2009 “Sickle Cell Disease Awareness and Education Strategy Development Workshop Report.” The report declares, “SCD is considered an ‘orphan disease’ (poorly publicized and not yet adopted by the medical research and funding communities) because it affects fewer than 200,000 people nationwide. Although the medical burden of orphan diseases is great, these diseases often are last to receive research funding.”²⁰ If that is true for SCD then why is it seemingly not true for Cystic Fibrosis?

The point here is that Cystic Fibrosis, like SCD, are both orphan diseases. The fact that SCD is viewed as an “orphan disease” by entities like the National Heart Lung and Blood Institute does not alter the fact that it has had devastating effects on members of humanity throughout the ages. It also does not justify the lower levels of research funding applied

towards finding a cure and improved treatment methods. In consideration of these facts, there should be no doubt that SCD has been the recipient of disparate treatment with regards to research. Untold millions of people have lived shortened, pain-filled lives because of SCD since at least the time of King Tut and millions more are bound for the same fate until a cure is found. Disparate treatment in research stands in the way of changing that.

Disparity in Government Support

A review of the 2012 Budget Justification for the Department of Health and Human Services (“HHS”) Centers for Disease Control and Prevention provides important material for the discussion about the disparate treatment of SCD. According to the justification, HHS is in the process of lumping all of its blood programs into one program called, “Public Health Approach to Blood Disorders.” The request explains, “Beginning in FY 2012, CDC will gradually transition its

four disease-specific blood disorder activities into one consolidated approach. The consolidated approach will broaden the work already being done through CDC's successful collaborations with a CDC-funded national network of 135 HTC's ["Hemophilia Treatment Centers"], national and community organizations, universities and other partners by providing flexibility to expand activities to include all non-malignant blood disorders with an immediate focus on disorders with the greatest burden and unmet need: DVT/PE, Sickle Cell Disease (SCD), and von Willebrand Disease. This broad public health approach to blood disorders will extend CDC's reach from approximately 20,000 people seen at HTCs to other patients with bleeding disorders currently treated outside the HTCs and the roughly four million people with one of the targeted blood disorders."²¹

The CDC's budget request for the Public Approach to Blood Disorders Program for 2012 was for \$20,165,000. However, the request does not clearly specify how much of that amount is dedicated to SCD. Although the justification does acknowledge its efforts towards SCD by stating that it had, "Initiated collection of data to describe epidemiologic and clinical characteristics of people with hemoglobinopathies through public health surveillance pilots on sickle cell disease and thalassemia,"²² it does not mention SCD in relation to any future activities. It does, however, state a goal that is of interest in our ongoing discussion of the disparate treatment between SCD and other orphan diseases. That goal is to, "Reduce the proportion of persons with hemophilia who develop decreased joint mobility due to bleeding into joints."²³ This leaves room for the question, could the CDC not also have a goal to reduce the proportion of persons with SCD who develop decreased joint mobility due to aseptic necrosis of the joints?

The U.S. government did have SCD legislation in place; the Sickle Cell Treatment Act of 2003 ("SCTA"). SCTA was the most comprehensive national SCD legislation since the Sickle Cell Anemia Control Act was signed into law in 1972. However, SCTA ran out of funding in 2009. In July 2011, Rep. Danny Davis (D-Illinois), offered a House Resolution (H.R. 2518) to provide the funding that SCDAA seeks for SCTA. H.R. 2518 would amend Section 712(c)(6) of the American Jobs Creation Act of 2004 (Public Law 108-357; 42 U.S.C. 300b-1) and would extend the authorization of appropriations for the Sickle Cell Disease Prevention and Treatment Demonstration Program for five (5) years.

In its original form, SCTA provided \$10 million in funding to create up to 40 Comprehensive Sickle Cell Centers around the country. That amount is \$7 million less and was intended to fund 95 fewer treatment centers than what was requested by the CDC in 2010 for Hemophilia. Since its introduction last year, H.R. 2518 has been sitting in the House Subcommittee on Health and no action has been taken on it since it was assigned to that committee on July 15, 2011. One does not have to look too far below the surface to see the disparity in how the nation treats these two disorders of the blood.

Representative Davis is not alone in his efforts to increase government support of SCD. Senator Benjamin Cardin of Maryland has also been a voice in the wilderness over the last several years. Speaking from the Senate floor on November 17, 2010, Senator Cardin said,

“The enormous human and financial cost of this disease underscores the importance of finding a safe cure for sickle cell disease. A worrying finding in research is that conscious or unconscious racial bias adversely affects the availability of resources for research, delivery of care, and improvement of that care. I am particularly concerned because there is a significant gap in funding for more publicized but less prevalent diseases as compared to sickle cell disease.”²⁴ Senator Cardin also said, “Despite increased research dollars for sickle cell disease and major advances in treatment, important gaps still exist in the equity of Federal funding allocation and in the provision of highly qualified clinical care.”²⁵

There is yet another government-based example of health disparity in regards to SCD and Hemophilia. The source of this disparity is what is known as the Federal 340B Program. According to the U.S. Department of Health and Human Services Health Resources and Services Administration website, “The 340B Drug Pricing Program resulted from enactment of Public Law 102-585, the Veterans Health Care Act of 1992, which is codified as Section 340B of the Public Health Service Act. The 340B Drug Pricing Program is managed by the Health Resources and Services Administration (HRSA) Office of Pharmacy Affairs (OPA). Section 340B limits the cost of covered outpatient drugs to certain federal grantees, federally-qualified health center look-alikes and qualified hospitals. Participation in the Program results in significant savings estimated to be 20% to 50% on the cost of pharmaceuticals for safety-net providers. The purpose of the 340B Program is to enable these entities to stretch scarce federal resources, reaching more eligible patients and providing more comprehensive services.”²⁶ Cancer, Black Lung Disease, HIV/AIDS and Hemophilia are the only medical conditions authorized for inclusion in the program. SCD is once again left on the outside looking in.

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Senator Benjamin Cardin

Although the savings available through the 340B are mostly available to the treatment centers and pharmacies participating in the program, it seems that SCD’s inclusion would allow for the same ability to “reach more eligible patients” and provide “more comprehensive services” to a much larger group of people that could use additional medical care and support.

In addition to the federal government, research of state supported Sickle Cell programs within the nation yields what appear to be some serious concerns regarding the treatment of the SCD population. It is acknowledged that there is a tremendous emphasis on new born screening at the state level. New born screening is very necessary but, in general, it represents the extent of most of the states’ support of Sickle Cell. Through new born screening, parents are able to learn if their child has Sickle Cell Disease or Sickle Cell Trait. However, only two of the 50 states within the union appear to provide some level of supportive services in addition to newborn screening.

The significant majority of care provided in those states is mainly available for children only. Certainly, the care of children with Sickle Cell or any other disorder is important but we must find a way to make the states recognize one very important reality; Sickle Cell does not go away once a child leaves adolescence. In fact, Sickle Cell has the tendency to get more vicious with every additional year of a person's life. Compounding this issue is the fact that Sickle Cell patients are living longer and, thus, placing greater demands on patients, families medical providers and community based SCD organizations.

Disparity in Health Care

“Obtaining specialty care can be a significant challenge as the number of health professionals trained to treat the disease is limited and the number of professionals specializing in the treatment of this disease is decreasing.”²⁷ This quote from the National Heart Lung and Blood

“The association between SCD-specific hospital volume and mortality also suggests that regionalization of SCD care, especially for adults, may have the potential to improve outcomes.”

Timothy McCavit, et al.

Institute's 2009 “Sickle Cell Disease Awareness and Education Strategy Development Workshop Report” succinctly describes a situation of great concern in the SCD field. Dr. Lauren Smith suggested something similar in her paper by stating, “Many primary care providers who practice in settings outside comprehensive sickle cell centers may not be fully aware of treatment guidelines. There is also a crucial need to increase the workforce capacity to care for adult patients with SCD to provide appropriate continuity of care for adolescents transitioning to adult care.”²⁸ Smith goes on to say, “However, there is no coordinated process to ensure the widespread adoption of treatment guidelines and no requirement for NHLBI-funded comprehensive sickle cell centers to track their implementation or assess their

effectiveness, such as occurs through the cystic fibrosis registry.”²⁹

In 2010, Timothy L. McCavit of The University of Texas Southwestern Medical Center and his team of experts published the results of a study in the American Journal of Hematology in which he said, “The association between SCD-specific hospital volume and mortality also suggests that regionalization of SCD care, especially for adults, may have the potential to improve outcomes.”³⁰ McCavit, et al, expounded further on this by saying, “Our findings suggest that outcomes could be improved for patients with SCD by identifying and highlighting SCD centers-of-excellence and through regionalization of SCD care. Primary care providers and hematologists for adult patients with SCD, in particular, might consider SCD-specific hospital volume when deciding where to admit or refer SCD patients.”³¹

As asserted in the aforementioned Education Strategy Development Workshop Report, “The best way to achieve optimal care for patients who have sickle cell disease, including preventive care, is for the patients to be treated in clinics specializing in the care of this disease. All sickle cell patients who have sickle cell disease should have a principal healthcare provider, and that provider, if not a hematologist, should be in frequent consultation with one.”³² Yet, for the

majority of SCD patients, such opportunities for specialized care are vastly out of reach. There are fewer than twenty (20) Comprehensive Sickle Cell Centers in the United States and, without reauthorization of the Sickle Cell Treatment and Control Act, or other similar legislation, the number of such centers will not be able to grow and may, in fact, decrease.

Currently, Comprehensive Sickle Cell Centers are located in Brooklyn, New York; Oakland, California; Pittsburgh, Pennsylvania; New York, New York; Greensboro, North Carolina; Chicago, Illinois; Philadelphia, Pennsylvania; Mobile, Alabama; Culver City, California; West Palm Beach, Florida; Hollywood, Florida; Memphis, Tennessee; Little Rock, Arkansas; St. Louis, Missouri; and Eastern North Carolina.³³ This means that many thousands of SCD patients living in other major population centers such as Detroit, Michigan; Dallas, Texas; San Antonio, Texas; Los Angeles, California; Phoenix, Arizona; Indianapolis, Indiana; and Washington, D.C. do not have the same level of access to the best available standards of care. It also means that there is a disparity within the SCD population itself. Those in areas near or adjacent to Comprehensive Sickle Cell Clinics have opportunities to access specialized care whereas those far removed from those centers do not.

It is only logical then that reductions in health care disparities for SCD patients can yield, not only fairness, but savings, as well.

There is yet another disparity to discuss; the availability of specialized treatment for children is much greater than it is for adults. The number one recommendation of the Health Care Discussion Group of the National Heart Lung and Blood Institute's ("NHLBI") 2009 "Workshop on Adults with Sickle Cell Disease: Meeting Unmet Needs" succinctly calls for a change to this paradigm. The NHLBI Health Care Discussion group's recommendation stated, "Multidisciplinary Comprehensive Center Model for children should be developed for adults. Centers should include community providers, community-based organizations, and researchers. Elements of care should include multidisciplinary teams, pain management, psychosocial programs, and day units."³⁴

Kauf pointed out that SCD health care costs exceed \$1 billion per year.³⁵ It is only logical then that reductions in health care disparities for SCD patients can yield, not only fairness, but savings, as well. Senator Cardin highlighted the importance of this fact in his address on the Senate floor in November 2010 when he said, "Besides our moral obligation to ensure that patients receive appropriate care, there is also an economic argument. Research showing the high proportion of sickle cell disease costs associated with inpatient hospitalization suggest that interventions that reduce complications such as pain crises could be cost-saving."³⁶

The consequences of disparity in health care treatment for SCD patients are gravely serious and this situation requires the nation's utmost attention. Experts tell us that increased access to better care will most likely result in improved quality of life for many who suffer from SCD. Consequently, the lack of availability of such care and the paucity of medical provider knowledge of recommended treatment protocols is more than just a disservice to SCD patients; it is, in fact, a form of appalling neglect. In other words, as Lauren Smith says, "It is tragic and

unjust for a particular group of patients to suffer avoidable complications and even death because effective new therapies have not been uniformly implemented. Although this is not unique to SCD, the severity of the disease and the nature of who suffers from it make the impact of this failure both severe and disparate.”³⁷ This is not something that we should tolerate as an advanced society.

Disparity in Health Care Insurance Coverage

The availability of affordable medical insurance is widespread in America, unless of course, one has a pre-existing condition like SCD. Even people with pre-existing conditions can acquire coverage but doing so may be either too expensive or the coverage may carry too many limitations. It is usually a combination of the two that restricts the availability of medical insurance for many SCD patients who do not have the luxury of being under an employer’s group plan. Some states like New Jersey, Alabama, Louisiana and Florida have recognized the

“In 2004, there were an estimated 113,098 hospital stays during which sickle cell disease (SCD) was noted, of which nearly three quarters were for adults with SCD.”

Claudia Steiner, et al.

importance of making private health insurance available for SCD patients and have passed laws that prohibit insurance carriers from denying coverage because of pre-existing conditions. Though this is a good step that many other states need to follow, the high cost of health care insurance or the inability to qualify for it remains an obstacle that many with SCD cannot surmount.

The Agency for Health Care Research and Policy released a report in December 2006 entitled, “Sickle Cell Disease Patients in U.S. Hospitals, 2004” as part of its “Healthcare Cost and Utilization Project.” The authors, Claudia A. Steiner and Jeffery L. Miller, provided the results of their

study of nationwide hospitalizations for SCD from 1994 through 2004. Their findings included staggering information about the number and frequency of SCD related hospitalizations. Steiner and Miller reported, “In 2004, there were an estimated 113,098 hospital stays during which sickle cell disease (SCD) was noted, of which nearly three quarters were for adults with SCD.”³⁸ The authors also stated, “The average cost for each hospitalization was \$6,223, and the total estimated cost for SCD hospitalizations in 2004 was \$488 million.”³⁹

In March 2012, the United States Government Accountability Office released a report to congressional requestors that provided statistical information about the utilization of private health insurance in America. This report was conducted to determine the impact that the Patient Protection and Affordable Care Act (“PPACA”) passed into law in 2011 would have on reducing the number of uninsured individuals who are denied coverage because of their pre-existing conditions. Since a provision of PPACA prohibits the exclusion of those with pre-existing conditions, the GAO report is helpful in reaching an understanding of the scope and impact of the pre-existing conditions exclusions. Among other things, the report found that between 36 and 122 million adults in the United States had some form of pre-existing condition.

The GAO report also revealed, “With certain exceptions, such individuals and any other individual attempting to purchase coverage in the private individual market can have coverage denied, offered at a higher-than-average premium, or offered with a rider that excludes coverage of a pre-existing condition. SCD was included as a pre-existing condition for the purposes of this report. The report found, “Adults with pre-existing conditions, on average, spend thousands of dollars more for all health care—between \$1,504 and \$4,844 more per year—than other adults.”⁴⁰

Steiner and Miller stated, “Among those principally hospitalized for SCD, more than three-quarters of stays were billed to public payers. Two-thirds of hospitalizations for SCD were billed to Medicaid, and an additional 13 percent were billed to Medicare. About 15 percent of hospitalizations for SCD were paid through private insurance, and 4 percent had no insurance.”⁴¹ Steiner and Miller also found that 34 percent (34%) of hospitalizations for all conditions were covered by private insurance. The GAO report stated, “Americans obtain health insurance coverage through a variety of private and public sources, but a majority—67 percent of adults as of 2010—rely on private insurance, most through employer-sponsored group coverage.”⁴²

In other words, there is a 30 percent to 63 percent (30% - 63%) difference in the utilization of private insurance coverage between those with SCD and those with all other conditions or those of the general population. Though there are factors that may impact the accuracy of this information, it is quite clear that SCD patients participate in private health care insurance at a disparate level.

The full implementation of PPACA in 2014 should have a dramatic impact on increasing the percentages of SCD patients with access to private health care insurance. However, the fact that this disparity exists at the present time should be enough to motivate society to address this problem. This is not as much about the numbers as it is about the lives of those with SCD. Numerous analyses, reports and papers have identified how crucial it is for people to have dependable health care coverage to improve their well-being and their quality of life. The same is true for those with SCD. In fact, it may be even more important for them to have access to regular medical examinations and treatments than for those who suffer from many other ailments.

Disparity in Social Service Support

The importance of psychosocial support for those living with SCD can never be understated. A statement in the National Heart Lung and Blood Institute’s (“NHLBI”) publication, “The Management of Sickle Cell Disease,” clearly identifies the importance of psychosocial intervention in the holistic care of the disease. It says, “The pain experienced by many patients with SCD can be demoralizing and overwhelming. In addition to the psychological effects of inadequately treated pain, patients have the added stress of continually searching for effective pain relief, resulting in frequent emergency room visits and episodic care. This cycle can lead to depression, which is highest among the chronically ill and in the 20-40 age group, and is often

not recognized or addressed. Continued comprehensive care - including a strong psychosocial component - for adults with SCD is most important, since prevention of complications is the key to longevity.”⁴³

A publication entitled, “Guidelines for the Treatment of People with Sickle Cell Disease,” written by members of the Sickle Cell Advisory Committee (“SCAC”) of the Genetic Network of New York, Puerto Rico and the Virgin Islands (“GENES”) with support from grants from the U.S. Health Resources and Services Administration echoes that of the NHLBI above. It states, “The initial bio-psychosocial diagnostic interview is critical in identifying such issues as the underlying genetic guilt parents may carry, as well as unresolved issues about their own trait status and its implications for future childbearing. Coordination of services demands an assortment of skills and abilities that are unique to social work and are provided by trained, licensed, professional social workers. Social workers coordinate services for patients and families with sickle cell disease by managing the systems that impact on the family homeostasis and equilibrium.”⁴⁴

“Continued comprehensive care - including a strong psychosocial component - for adults with SCD is most important, since prevention of complications is the key to longevity.”

National Heart Lung and
Blood Institute

One of the primary reasons that psychosocial support is so important is that its effective utilization can minimize the amount of stress that SCD patients must cope with. The existence of too much stress is often responsible for the onset of pain attacks. That is why SCAC’s guidelines pronounce, “Without social work serving as the “hub” of comprehensive care services and networking for patients and families, both quality of life and longevity are jeopardized. Social workers intervene to minimize the threat of family breakdown and disequilibrium.”⁴⁵ Statements similar to those of NHLBI and SCAC can be found in numerous articles and literature concerning SCD, thus validating the importance of psychosocial support in the management of the disease.

Just how accessible is psychosocial support for SCD patients? Does the availability of this type of assistance exist in a manner that is equal to that of other chronic diseases? The answer to that question is difficult to ascertain but, interestingly enough, a study conducted by Donna K McClish, et al, reveals a hint. McClish found that SCD patients “reported a HRQOL (“Health Related Quality of Life”) that was equal to or poorer than patients with other significant chronic conditions in many domains. Even more interesting is that McClish’s work revealed that people living with Cystic Fibrosis demonstrated a better HRQOL than those with SCD. She states, “Similar to patients with SCD, until somewhat recently, patients with cystic fibrosis rarely lived until adulthood, marking this as a disease with significant sequelae and high mortality. It is interesting, then, to see that quality of life of adult survivors of this chronic disease, even though impaired, was comparable to national norms, and was generally far superior than that reported by adults with SCD.”⁴⁶ Once again, we see that comparatively speaking, Cystic Fibrosis appears to fair better than SCD.

Disparity in Advocacy

In her paper, Smith identifies yet another area where SCD receives disparate treatment. She states, “For example, for fiscal year 2003, the Sickle Cell Disease Association of America’s total revenue was \$498,577 compared with \$152 million for the Cystic Fibrosis Foundation, a 300-fold difference that has substantial implications for the Sickle Cell Disease Association of America’s ability to support research and advocacy.”⁴⁷ SCDA’s budget has grown since 2003 but it has not grown as much as that of the Cystic Fibrosis Foundation. Guidestar.org, an online service that provides information about 1.9 million charities, lists SCDA’s 2010 revenue at \$1,273,031.⁴⁸ In contrast, Guidestar reports that the Cystic Fibrosis Foundation had revenues of \$313,308,873 for the same year.⁴⁹

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Yet another example of inequality is uncovered when looking at the 2011 financial statement for the National Hemophilia Foundation (“NHF”). That report shows that NHF had revenues of \$11,026,142 and had \$6.9 million in cash and equivalents at year’s end.⁵⁰ Here is yet another illustration of disparity. There are approximately 30,000 people in the United States with Huntington’s Disease, a genetically transmitted brain disorder. As is the case with Hemophilia, there are fewer than one-third the number of people with Huntington’s Disease than there are with SCD. Yet Guidestar reports that the Huntington’s Disease Society of America (“HDSA”) operates on revenues of \$8.6 million, or nearly seven (7) times more than that of the SCDA.⁵¹ Even the National Tay-Sachs and Allied Diseases

Association (“NTSAD”) which supports people with the very rare Tay-Sachs Disease had proportionately higher revenues than those of SCDA. Approximately one in every 27 Jews in the United States of America is a carrier of the Tay-Sachs gene as opposed to one in every 12 African-American carriers of the SCD gene. NTSAD had revenues of \$892,679 in 2010.⁵² This is just a few hundred thousand dollars less than the revenues for SCDA, an organization that supports the needs of a population that is more than twice as large.

The above examples are reflective of the overall pattern of disparity between how SCD and other genetic disorders are treated in America. The availability of revenue is paramount to a charity’s ability to get its message out to the public. A greater availability of revenue makes it possible for these and other organizations to do a much better job of advocating for their respective causes. The fact that SCDA operates with a fraction of the funds available to other charitable organizations serving significantly smaller populations with genetic disorders means that it does not have equal ability to spread its message to generate support for its cause.

Discussion

There are a number of factors contributing to the long-running lack of regard that is afforded to SCD. Chief among these factors is the racial bias that “adversely affects the availability of resources not only for research and the delivery of care, but also for improvement of that care,” referenced by Dr. Lauren Smith and others cited in this paper.

As stated previously, only \$1 million had been spent on SCD research in the first sixty-two (62) years after its discovery in 1910. We are all well aware that racism was a fact of life in America and throughout the world during that shameful period of time. During that era, society mistakenly believed that SCD only affected people of African descent. People of African descent were regarded as subhuman, openly mocked and humiliated, generally despised and even murdered for looking at someone the wrong way. Since Sickle Cell only affected the “coloreds” and, especially since it was known that it is not contagious, society had no motivation to find a cure. This same pattern of bigotry has negatively impacted potential advancements in SCD research and care as much as, if not more than, it has impacted the availability of equal opportunities in employment and housing.

Another of the factors is the relative level of indifference displayed by the SCD community over the years. This indifference is actually more like apathy. Apathy is defined by Webster as “lack of interest or concern.” From all intents and purposes, it appears that a lack of interest or concern has been a major obstacle towards finding a cure for Sickle Cell and for creating adequate funding to support the 100,000 or so Americans, and millions worldwide, who suffer from it.

We must ask ourselves these questions: Why is it, in 2012, that Sickle Cell still exists without a cure? Why is it that our social service networks in 48 of our 50 states still don't have any kind of support net for people with Sickle Cell, especially adults? Why is it that 36 meritorious recommendations from the 2002 “Workshop on Adults with Sickle Cell Disease” are still just recommendations?”

The answer to all of these questions is the same; because that is what has been allowed. Now that it is understood that SCD affects a wide variety of ethnicities and that people are living longer with it, isn't it time that we seek a change in how society deals with it?

The great Fredrick Douglas once said, “Power concedes nothing without a demand. It never did and it never will. Find out just what any people will quietly submit to and you have the exact measure of the injustice and wrong which will be imposed on them.”⁵³ Throughout history, improvements in the human condition have arisen only as a result of the people's demands for those improvements. Where are the demands for a cure for Sickle Cell? Where are the demands for more supportive services for people who have to deal with the uncontrollable, unbearable and unending pain?

In the field of behavioral psychology, it is commonly accepted that one must honestly acknowledge that a problem exists if they are to have any chance of successfully treating and correcting it. It is imperative, therefore, that society acknowledges that it has discriminated against SCD since its discovery and that it is on track to continue doing so unless this pattern of injustice is broken. Only then can we move forward with correcting our societal indifference towards SCD and only then can we start to witness advancements that allow us to more effectively meet the unmet needs experienced by its sufferers.

Recommendations

The following recommendations are presented for the purpose of providing guidance for the rectification of the current disparities that exist in the areas identified in this paper. These recommendations should receive all due consideration to the potential positive impact that their implementation would have on the lives of the people who suffer from one of nature's most commonly occurring genetic disease.

Recommendation #1: CDC Adoption of SCD as a Health Disparity Topic

First and foremost, it is recommended that the Centers for Disease Control, in concert with the U.S. Department of Health and Human Services, declare SCD to be a health disparity. Such an endorsement would encourage a higher level of awareness and it would ultimately lead to improved health care and access to psychosocial services for those afflicted with SCD by providing the necessary validation for all relevant entities to take SCD more seriously in the coming years.

Recommendation #2: National Institute of Health Adoption of SCD as a Health Disparity

It is recommended that the National Institute of Health ("NIH") adopt SCD as a health disparity. NIH is the largest source of funding for medical research in the world. Its mission is to seek fundamental knowledge about the nature and behavior of living systems and the application of that knowledge to enhance health, lengthen life, and reduce the burdens of illness and disability. The National Institute on Minority Health and Health Disparities ("NIMHD") within the National Institute of Health is responsible for the development of priorities, objectives, budgets, and policy statements for the conduct and support of all NIH minority health and health disparities research activities. It develops and maintains a Health Disparities Information ("HDI") database to facilitate the collection of data, translation of research, education, dissemination, and communication of information to various audiences. It also develops and revises, as necessary, the national definition for health disparity population in consultation with the Agency for Healthcare Research and Quality. The adoption of SCD as a health disparity by the NIH will facilitate more equal treatment in the allocation of research dollars.

Recommendation #3: Federal Legislation to Ensure Equality in Genetic Disease Support

There is a myriad variety of state statutes that provide different levels of support for different diseases. Additionally, the federal government itself is perpetuating the disparate treatment of diseases through its provision of the 340B program that provides prescription medication assistance for people with Hemophilia but not SCD, Cystic Fibrosis and other diseases that require significant amounts of prescribed medications for health maintenance. Therefore, it is recommended that the members of Congress recognize, acknowledge and correct these disparities to ensure equal levels of support for persons with genetically transmitted disorders at both the federal and state levels.

Recommendation #4: Reauthorization of the Sickle Cell Treatment Act of 2003

The Sickle Cell Treatment Act of 2003 (“SCTA”) ran out of funding in 2009. SCTA is the most comprehensive national SCD legislation since the Sickle Cell Anemia Control Act was signed into law in 1972. Among other things, SCTA authorized and funded the implementation of Comprehensive Sickle Cell Centers (“CSC”) around the country. There are currently about 141 federally-funded hemophilia treatment centers and programs in the United States. In contrast, there are only 15 Comprehensive Sickle Cell Centers. It is recommended that SCTA be reauthorized and sufficiently funded so that significantly more CSCs can be established to more equally serve a population that is significantly larger than that of hemophilia.

Recommendation #5: Careful Monitoring of Insurance Company Practices After PPACA Implementation

When implemented in 2014, the Patient Protection and Affordable Care Act is supposed to ensure that no person is denied health care insurance coverage or penalized for a pre-existing condition. In that SCD patients have historically been among those discriminated against by being denied coverage or being forced to pay increased premiums, it is recommended that watchful government eyes be put in place to ensure that such barriers are forever eliminated. The impact of this will be improved access to and utilization of health care and, subsequently, an improved health related quality of life and a decreased burden on Medicaid and Medicare.

Recommendation #6: Eliminate Disparities in Psychosocial Services at the State Government Level

It is recommended that state legislatures across the country establish legislative study committees to review and analyze the disparities in the existence of federal or state funded programs that offer any form of psychosocial services for genetically transmitted disease patients. These committees should then work with their respective legislative bodies to create and pass legislation that removes any existing disparities within their statutes, rules and administrative codes.

Recommendation #7: Increase Activism and Advocacy within the SCD Community

It is not solely the fault of the government or society as a whole that organizations supporting those with SCD are so vastly underfunded. The more well-funded genetic disease organizations are in that position because those affected have been more vocal and active in their causes, they have advocated incessantly and have done so with passionate fervor. Therefore, it is recommended that anyone and everyone affected by SCD immediately begin to rally behind their cause with spirited enthusiasm and intensity by advocating for, donating to and volunteering at all SCD organizations at levels like those never witnessed before.

Conclusion

As a disease that primarily affects persons of color, and in knowing the history of racism in America and around the world, it is somewhat understandable that SCD has received disparate treatment by society. Although the cited disparities may be understandable in that context, they are by no means acceptable. The disparate treatment of individuals for any reason should not be tolerated but it is particularly regretful that unequal treatment exists for individuals who bear no personal responsibility for having a disease like SCD.

This paper has demonstrated that disparate treatment of SCD exists in regards to inadequate recognition by the Centers for Disease Control, its status as an underfunded focus of research activities, the unbalanced support it receives from government and the unequal availability of and access to health care by its sufferers. In addition, evidence for the disparate treatment of SCD is present in the availability of and access to health care insurance, social service support and advocacy.

A nation of healthier people equates to a nation with lower health care costs and greater productivity. Therefore, the elimination of any of the disparities identified in this paper will yield improvements not only in the health related quality of life of SCD patients; it will also provide enhancements to the social and economic well-being of the nation. A series of recommendations have been offered in this paper to facilitate those improvements. These recommendations range from elevating the status of SCD so that it becomes a recognized health disparity at the CDC to calling for expanded activism and advocacy within the SCD community.

It is my hope that this paper can help raise awareness about these disparities and that those with SCD will soon begin to receive equal treatment.

About the Author

Gary A. Gibson is the President and Chief Executive Officer of Martin Center, Inc. in Indianapolis, Indiana. Established in 1969, Martin Center is one of the nation's oldest 501(c)(3) Sickle Cell Disease supportive services agencies. Mr. Gibson has served in various high-level Indiana state government positions and is a two-time recipient of Indiana's Sagamore of the Wabash Award, the state's highest honor for civilians. He was married to a Sickle Cell patient for nearly twelve years before her death at the age of 36 in December 1989. Mr. Gibson later married the woman that was once his high school sweetheart and he resides happily with her in Indianapolis, Indiana. He has also authored another white paper about SCD, "Sickle Cell Disease: Still Here and Still Causing Pain."

End Notes

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