Sickle Cell Disease: 
Still Here and Still Causing Pain

A Broader View 
of the Impact and Needs of SCD

Introduction

Chances are that you might not have heard much about Sickle Cell Disease ("SCD") lately. That doesn't mean that it has gone away. It certainly hasn't. SCD is still negatively impacting the lives of millions of people worldwide. There are as many as 150,000 babies born with the disease each year in Nigeria, alone.\(^1\) Closer to home, there are an estimated 72,000 to 100,000 people\(^2\) living with the disease in the United States, with over 1,000\(^3\) of them living in the state of Indiana. That means that there are over 1,000 Indiana citizens living in pain through no fault of their own. It is not their fault because SCD is a genetic disease, not one that is acquired through negative behavioral choices.

SCD is despicable and the way that it has been handled by society is even more
contemptible. It is the most common of humanity’s genetic disorders and it is nothing less than appalling that it still exists. There is too little money to find a cure, there is too much apathy about it (including within the minority populations that it impacts the most) and there is too little financial and psychosocial assistance available to help the adolescents and adults who must carry its pain-filled burden each and every day.

This is not to say the SCD does not receive attention but rather that it does not receive the level of attentiveness that it deserves. The nature of SCD is such that it causes overburdening problems in the lives of those who have it and its very existence presents wide-scale problems that affect both individual quality of life and the economic well-being of humanity as a whole. The truth of the latter statement can be found in the combined untold value of lost human productivity and the ever-increasing expenses related to the medical treatments for SCD patients.

Of course, the hallmark feature of SCD is the pain that it causes. Most people who know anything at all about SCD know about the intense physical pain that occurs during what’s known as a “Sickle Cell Crisis.” However, fewer people are aware of the other forms of pain that SCD generates in the lives of those who must live with it. Included in these other forms of pain are the psychological, social and financial struggles that are associated with any chronic disease.

In some parts of the world, particularly Africa and India, SCD is seen as a curse and people who have it are ostracized from their tribal societies. A certain amount of that belief still persists, albeit to a lesser degree, in other more advanced societies around the world. Then there is the psychological burden of living a life that can be interrupted at any time, without warning, by the onset of severe pain that can persist for days on end.

In addition to the psychological pain, SCD leads to social pain, specifically the kind of pain that results from the inability to socialize to the same degree that most healthy people enjoy. SCD is responsible for missed social events and the sometimes lengthy periods of isolation resulting from the recurrences of crises that can impede participation in social activities. Furthermore, SCD interferes with romantic activity and it continuously complicates family dynamics in a varying number of ways.

SCD also produces economic pain. It requires extensive access to and utilization of medical services, equipment and prescription medications. Consequently, many SCD patients and their families find themselves confronted with exorbitant medical bills. In many cases, employment is difficult to find and/or maintain. This creates high-stress cash flow management issues that bring their own form of agony.

This paper will encourage a renewed emphasis on increasing not only the awareness of SCD itself but also on broadening public understanding of the many needs of SCD patients that are going unanswered. Of particular emphasis
are the needs of adolescents and adults who now live well beyond the life expectancies that were common in the middle part of the 20th century. Not long ago, SCD patients were expected to live no longer than 20 years. Improvements in diagnosis and treatment have generally doubled the life spans of SCD’s sufferers. That is important and commendable progress but life expectancies of 42 – 48 years are nothing to be proud of in today’s day and age, especially in developed nations. Regardless of this, the fact remains that there are many thousands of people in the United States and millions worldwide who are not receiving adequate support to bolster their quality of life. The impact of SCD is much greater than most people realize. This must be changed for humanity’s sake.

A Description of SCD

SCD is caused by an abnormal type of hemoglobin called hemoglobin S. Hemoglobin S changes the shape of red blood cells, especially when the cells are exposed to low oxygen levels. As the oxygen in the red blood cells depletes, they become brittle and break into shapes that resemble crescents. These sickle-shaped cells deliver less oxygen to the body’s tissues, organs and bones. The sickled cells lodge in small blood vessels and interrupt the flow of blood to any given area of the body. When this happens, the SCD patient is said to be in “crisis.” In addition to the sometimes prolonged periods of intense pain, the long-term results of SCD can be necrosis, organ damage, joint damage, various medical complications and shortened life spans.

SCD crises, which can last for days or weeks at a time, often require hospitalization and the administration of narcotic pain killers, hydration and blood transfusions. In addition to the suffering caused by the pain and the inconvenience of frequent hospitalizations, SCD families often find themselves overburdened with medical expenses. Statistics from the Center for Disease Control indicate that the annual cost of hospital treatments for children with SCD can be as much as $13,000 higher than that of children who are generally healthy. The CDC also reports that between the years 1999 and 2007, there was an average of approximately 197,333 SCD related emergency room visits in the United States per year.

Statistics like these shed a new light on the problem that is SCD and they make it easier to understand that even though it affects a seemingly small percentage of the general population, SCD is a giant of a disease that creates an enormous footprint on people’s lives and on our health care system.

The People with SCD

One of the difficult challenges of SCD is that it affects individuals in so many different ways. Take Peter for example. Peter is 21 years old and has already had five strokes. SCD has tried it’s best to keep him from realizing his dream of higher education, but as a result of his perseverance and the help of his family and others, he recently graduated from high school. Then there’s Kellen. In
most ways, Kellen is just like other nine year old boys. He has a bright outlook on life and he has big dreams for a successful future. However, SCD constantly reminds Kellen that he is different from most of the other boys that he knows. After repeatedly sending him to the hospital and even causing him to lose his spleen at such a tender young age, SCD has not defeated Kellen or his loving family. He still goes to school and still holds onto his dreams. There are many, many others like Peter and Kellen who are quietly battling SCD on a daily basis.

SCD’s victims are of many ages and, though most people may not know it, they are of many ethnicities as well. For decades in the United States, it was erroneously believed that SCD was a “Black persons” disease, affecting only people of African ancestry. We now know that nothing could be further from the truth. SCD is the most commonly occurring genetic disease in the world. It can and does affect people from Latin America, the Middle East, India, Asia, Italy and Greece. In fact, it has been proven that SCD can exist in any area of the world where Malaria is, or has been, present. Also, the global rise in the number of inter-racial marriages and relationships is making it possible for the Sickle Cell Trait to pass to virtually anyone, regardless of the color of their skin. People with Sickle Cell Trait carry only one copy of the altered hemoglobin gene and generally do not experience the symptoms typically associated with the disease. However, when both parents carry the Sickle Cell Trait, there is a twenty-five percent (25%) chance that one or more of their children will be born with SCD.

It is interesting to note that one of the most famous personalities of all time, Egypt’s King Tut, may have died from SCD complications at the early age of 19 years. In a letter to the Journal of the American Medical Association published in June 2010, two German scientists stated that forensic evidence indicated a strong probability that the young king suffered from SCD and likely died as a result of its complications. The researchers state that SCD is the most common cause of bone damage like that observed in King Tut's mummified remains. They also suggest that since Tut's parents are thought to be related, there is a strong chance that they both carried the Sickle-Cell Trait.

**SCD Research**

From the time of its identification in 1910 by Dr. James Herrick until the Civil Rights era of the 1960s and 70s, SCD received very little attention and even less funding for research. In a 1970 article published in the Journal of American Medicine, Dr. Robert B. Scott stated, “In 1967 there were an estimated 1,155 new cases of SCA [Sickle Cell Anemia], 1,206 of cystic fibrosis, 813 of muscular dystrophy, and 350 of phenylketonuria. Yet volunteer organizations raised $1.9 million for cystic fibrosis, $7.9 million for muscular dystrophy, but less than $100,000 for SCA. National Institutes of Health grants for many less common hereditary illnesses exceed those for SCA.” Dr.
Scott’s article is widely credited with paving the way for the passage of the National Sickle Cell Anemia Control Act (“NSCACA”) in 1972.

Although he was factually incorrect in making the statement, “This disease is especially pernicious in that it strikes only Blacks and no one else,” President Richard Nixon signed the bill into law on May 16, 1972. In addition to the establishment of screening and education programs, NSCACA directed that more federal funding ($10 million) be spent on SCD research. It also led to the enactments of multiple SCD focused state laws and programs around the nation during the early 1970s.

Yet, even four decades later, the amount of money spent on SCD research pales in comparison to that of other diseases that do not seem to discriminate on the basis of ethnicity. According to data from the National Institute of Health, an approximate $73 million will be spent on SCD Research in 2011. To be fair, SCD research spending levels are higher than some other commonly known diseases like Psoriasis ($13 million), Spina Bifida ($12 million) and Uterine Cancer ($26 million). However, in contrast, research funding for Alzheimer’s Disease will be at $450 million; Asthma research will total $244 million; Cystic Fibrosis will reach $86 million; Epilepsy research spending will be around $134 million; and Lupus research will top out over $110 million. These statistics clearly demonstrate the funding inequity that SCD research has had to endure. The statistics also show that more research funds need to become available to expedite progress toward finding a cure and better treatment protocols for those suffering from SCD.

The Economics of SCD

There is much more that needs to be done to support those who have SCD. Among the most important areas to address is that of economics, for SCD causes tremendous economic hardships for patients and families. Due to its cruel behavioral characteristics including chronic pain, physical debilitation and emotional distress, SCD prevents many otherwise productive citizens from being able to acquire and maintain employment. Many SCD patients live at or below the poverty level. Consequently, many of them qualify for and receive Medicaid and Social Security Benefits. Though these benefits are helpful, they only marginally enhance the quality of life for those who have SCD. Many SCD patients are eager to make something of their lives and they are actually ashamed of the need to receive government financial assistance. We need to find a way to help them use their talents to contribute to society, not take from it.

One particular SCD patient, a young lady named Bianca, has just graduated from high school and wants to start working. If she is able to find employment, she will no longer qualify for Medicaid assistance. This poses a great dilemma for Bianca and her single father since the cost of her medications is approximately $9,000 per month. This is an expense that neither Bianca nor her
family can afford. Also, it is unlikely that any new job she finds will have a benefits program in place that would mitigate the high cost of the medicine that she needs to stay as healthy as possible. Consequently, it appears that Bianca’s entry into the workforce would cause her to suffer tremendous economic hardship which becomes a disincentive for her to realize her dream of working to support herself.

Bianca is certainly not alone. Her situation is indicative of a serious systemic problem that impacts a large and growing number of SCD patients and their families.

The economic ramifications of SCD are staggering. It also places an extremely heavy burden on the nation’s health care system. 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.\(^\text{10}\)

Kauf’s team analyzed data from the Florida Medicaid program on 4,294 SCD patients and then estimated the national cost of medical care based on 70,000 individuals with SCD. The researcher’s estimate of 70,000 individuals living with SCD is low by most calculations. That means that the actual annual cost of SCD medical care is quite possibly much higher than their $1.1 billion estimate. Kauf and her team stated that “Despite reductions in morbidity and mortality associated with early screening, sickle cell disease (SCD) is still marked by high utilization of medical resources.” Her findings include the statement that “the average cost per patient-month was $1,389.” The study also reported, “For an average patient with SCD reaching age 45, total undiscounted health care costs were estimated to reach $953,640. Overall, 51.8% of care was directly related to SCD, the majority of which (80.5%) was associated with inpatient hospitalizations.” Kauf’s report directly addresses the main premise of this paper. She states, “When one considers the additional contributions of SCD associated with reduced quality of life, uncompensated care, lost productivity, and premature mortality, the full burden of SCD is likely to be quite higher than the figures reported here.”

So who is paying for all this medical care? A paper published by the Connecticut Department of Health published in December 2007\(^\text{11}\) provides the answer. Data presented in this paper show that, in Connecticut, Medicaid paid for 47% of all discharges while Medicare paid for 24%. Private paid party payers (indemnity, managed care) paid a combined 24%. In other words, nearly three-quarters of SCD’s medical costs are borne by the U.S. government.

This fact alone should be enough incentive for the nation to find a cure for this despicable disease. Currently, that cure remains elusive. In the meantime, SCD is still causing physical, psychological, social and financial pain for tens of thousands of Americans and millions of others worldwide.
The Current Toll and Future Demands of SCD on Society

A 2006 World Health Organization’s report states, “The availability of diagnosis and treatment inevitably leads to a cumulative increase in numbers needing care, as patients live longer. The other usual consequence is an increase in annual cost per patient, which can have serious implications on countries, especially those with limited resources.”\textsuperscript{12} The report further expounds on this issue by stating, “As with all chronic disorders, improved management creates a cumulative demand for more services. Surveillance and education must be delivered at the community level through the primary health-care system so as to increase public awareness of the problem and lengthen the survival of affected individuals.”\textsuperscript{13}

Dr. Kofi Anie of the Brent Sickle Cell and Thalassaemia Centre in London identifies why the increasing life expectancy of patients with SCD is actually problematic in some respects. Dr. Anie states, “Psychological complications in patients with SCD are common. These range from inappropriate coping strategies, reduced health-related quality of life as a result of negative mood, and daily activity and role limitations, to neurocognitive impairment.”\textsuperscript{14} Dr. Anie concludes, “In the absence of a universal cure, it is recommended that psychological interventions should be incorporated into protocols for the management of patients with SCD and offered as standard care to help improve their general quality of life.” The implications here are simple and straightforward. More people living longer with SCD will require more of the psychological interventions Dr. Anie mentions.

The effects of SCD have life-long implications. In a feature article of the “Journal of School Nursing,” authors Sara Day and Elizabeth Chismark state, “The effects of these complications may lead to academic failure, limited career options, and for some, total disability. Despite studies describing the significant academic and cognitive impact of sickle cell disease, reports describing interventions are limited. There is a lack of awareness among educators of the academic risks associated with sickle cell disease and a lack of appropriate resource allocation.”\textsuperscript{15} Day’s conclusions are both valid and alarming for anyone who cares about education, self-empowerment and societal, cultural and economic progress.

Day’s suppositions are important because they correspond with the research conclusions drawn by a team led by Robert Noll. Noll’s team states, “Overall, these data suggest that despite the occurrence of an especially challenging disease, the social functioning of children with SCD who have not had an overt stroke is not impaired.”\textsuperscript{16} In other words, SCD does not prohibit its sufferers from having the ability to participate socially with other members of their communities. However, there are significant challenges standing in the way of Sickle Cell patients who are desirous of contributing to the economic and social well-being of their communities.
According to a team of researchers under the leadership of Julie A. Panepinto, “The impact of sickle cell disease on the HRQL [Health-Related Quality Life] of children is apparent, despite whether the children are living in homes with the lowest family income. Because many of these children do live in poverty and have other medical or neurobehavioral co-morbidities in addition to sickle cell disease, the impact of sickle cell disease on HRQL is even more severe.” Although the focus of Panepinto’s research was on children with SCD, there should be no arguments in the inference herein that her findings are also applicable to adults. In fact, such an inference is necessary and is predicated on the fact that there is a void in the availability of similar studies with an adult-oriented focus.

Adults with SCD are quite possibly one of the most neglected groups on Earth. Very little research has been conducted on their unique issues and even less psychological, financial and social assistance is available for them. The result is a tragic lack of understanding of their plights and a general absence of support mechanisms for them. The fact is that, once SCD patients pass beyond pediatric age, they no longer have access to the same level of comprehensive health and social services that they were accustomed to receiving as a child. They then fall into a lonely vacuum that is exceedingly difficult to fill. It is a void from which many of them never escape. Adults with SCD have been forgotten and, in many cases, they are left alone to shoulder their very heavy burdens. Society is treating these individuals poorly and they have very little means to change this situation on their own. If there was ever a group for which impassioned advocacy is needed, adults with SCD must surely rank high on the list.

One of the few studies available in the area of the socioeconomic impact of SCD on adults was written by Shawn M. Bediako. His findings include a straightforward assessment of the importance of this topic. Bediako states, “Adults with SCD constitute an emerging vulnerable and underserved population, and these findings suggest that there is a need for additional research on employment status and vocational outcomes in this group. While employment may not be identified as an area of primary concern for researchers or those who provide health care services to adults with SCD, it is important to note that in this era of health care reform, having a job serves as a gateway for access to health care services for many people. Thus, the ability to maintain gainful employment is critical for SCD patients and may be related to a host of important outcomes, including pain management behaviors, health care utilization, mental health, quality of life, and health care expenditures.”

The future demands of SCD on society are many. As more SCD patients live longer, more demands will be placed on those few systems, institutions and organizations that provide supportive services to them. In addition, the ever changing societal view of interracial marriages
The current explosion of immigrant populations in developed countries like the United States requires particular consideration, as well. Many of the immigrants have ancestral ties to areas of the world where SCD is prevalent and many of them may be carriers of the Trait and may have never been screened for it. Findings of the 2010 United States Census support this concern. According to the U.S Census Bureau, “The Hispanic population increased by 15.2 million between 2000 and 2010, accounting for over half of the 27.3 million increase in the total population of the United States.” The Census also states, “The Asian population alone increased by 43 percent between 2000 and 2010, more than any other major race group.”

Hispanic and Asian ethnic groups are documented carriers of SCD and the SCD Trait. In fact, 1 in 1,400 Hispanics and as many as 1 in 725 East Indians have SCD. As more unscreened immigrants produce offspring amongst themselves and with members of other ethnic groups, the probability for spreading the SCD Trait and SCD is certain to rise within the United States and other developed nations like the United Kingdom and France. Consequently, it is imperative that these individuals receive education and screening as early as possible upon their entry into these countries. Therefore, aggressive outreach, education and voluntary testing efforts are needed to prohibit the increase of SCD among the populace.

**SCD Needs and Solutions**

Providing for the current and future needs of SCD patients will continue to be a challenge, especially in consideration of the projected continuance of the low levels of funding that have been available to provide supportive services for them. As stated previously, there are many needs in the realm of SCD and its patients. Some of the most urgent needs are as follows:

**SCD Need: Health Care Provider Education**

Due to the nature of the disease, which includes the many ancillary complications, access to high quality health care is essential for the patient with SCD. Yet, even more than 100 years after its discovery, there is still a significant amount of medical inexperience regarding the treatment and patient management protocols for SCD. There are still many doctors and nurses who have never treated a SCD patient. This is understandable to a point because of the low number of people who have SCD but it is not good for the patient who ventures into an emergency room or neighborhood clinic while in the middle of a Crisis. A recent report published by the U.S. Department of Health and Human Services springing from the “Sickle Cell Disease Awareness and Education Strategy
SCD patients are not only at the mercy of the disease, they are also at the mercy of the health care providers who treat them. This is especially true since many SCD patients will need to seek medical care numerous times each year. The lack of knowledge of SCD treatment protocols is to blame for inadequate treatment of symptoms, erroneous decisions about when to release patients from medical care and, in some cases, avoidable and premature death. This situation should not exist but it does. The need to improve medical provider knowledge must be addressed with vigor but doing so will require significantly more resources than are currently available.

The fact that many health care providers are unfamiliar with SCD causes some of them to wrongfully label SCD patients as drug addicts. SCD patients sometimes have to endure this form of stigmatic profiling that is associated with their requests for narcotic pain relievers. In general, SCD patients do not like to take narcotics but the intensity of their pain can causes them to need something stronger than the over-the-counter pain relievers that are available. Health care providers must become more sensitive to the needs of their SCD patients and they must become more understanding of the acute and relentless levels of pain that arise during a crisis.

**Solution:** Establish and sufficiently fund a National Sickle Cell Disease Medical Provider Education Program within the United States Department of Health and Human Services that will provide federal grants directly to qualified SCD support agencies for the purpose of improving the SCD education level of medical providers.

SCD Need: Social Service Assistance

As stated, tremendous strides have been made in the diagnosis and medical treatment of SCD. What is lacking is the same type of advancement in addressing the psychosocial needs of those affected by the disease. This situation is briefly but eloquently stated in a briefing paper published by the United Kingdom’s Race Equality Foundation. Although the paper uses data and research that primarily originates in the U.K., the validity of its findings clearly apply on a global scale. The authors, Karl Atkin and Elizabeth N. Anionwu state, “Despite improvements in clinical management, poor service support remains a long-standing difficulty. The problem reflects a broader failure to engage with the social consequences of the conditions.” Atkin and Anionwu also state, “Service coordination, evidence-based practice, inter-agency collaboration and strategic leadership represent key challenges, as does the need to provide equitable care, wherever a person might live.”
A legislative task force on SCD in the state of Arkansas found what many who work in the field already knew. The report states, “As is the case with other chronic conditions, persons with sickle cell disease may need supportive counseling or therapy. Often they find their therapists do not understand their disease process, which makes it difficult for the patient to find the help they need. Many also have difficulty paying for those services.”

The task force report also includes the statement, “The support groups in the state receive numerous requests for assistance with an array of financial needs. Those community-based organizations have resources to assist with only a small fraction of the requests they receive.”

Unfortunately, what is true in Arkansas is true in every U.S. state and, of course, around the world, as well.

Lynne D. Ray, R.N., Ph.D., is an Associate Professor at the University of Alberta Canada. She studied 30 caregiving families and published an informative paper in 2003 in the Journal of Family Nursing. Ray said, “An important goal of special-needs parenting is to help one’s child secure a place in society. It is societal perceptions of disability and dependency that makes this goal particularly challenging for parents.”

Although Ray did not specifically cite SCD in her research, her findings indicate that the social systems in place to help those dealing with chronic disease were not always able to address their special needs. This was especially true for rare conditions. Ray’s paper states, “Children who had a common diagnosis were at a distinct advantage, because clinics, programs, and subspecialists were in place to serve their population group. To have a rare diagnosis, or worse yet no diagnosis, meant that services were much harder to find.” She added, “Although parents were intimately aware of numerous constraining system conditions, they doubted that professionals were aware of the consequences that these system structures or policies had for families.”

Most people within the SCD community are well aware of the systemic shortcomings that prohibit adequate psychosocial support for those with the disease. Yet, for various reasons, including the varying effects of SCD on different individuals who have it, it has been difficult to substantially address their needs on an aggregate basis. Nevertheless, it is imperative that we continue to make progress on identifying and addressing the social needs of those with SCD. Since it is true that SCD patients are living longer lives, it is also true that the demand on the institutions that treat and provide services to them is increasing.

In the case of SCD, longer life expectancy does not automatically equate to improved quality of life. What this means is that SCD patients will have longer lives filled with pain and suffering and all of the other aspects of the disease that manifest themselves in so many ways. In other words, more psychosocial help, not less, will be needed in the years to come. We must be ready, willing and
able to provide the support that is needed. Not doing so is not only a disservice to those who did not ask to have the disease; it is a disservice to those with whom they share their lives.

Solution: Establish and sufficiently fund a National Sickle Cell Disease Social Services Support Program within the United States Department of Health and Human Services that will provide federal social service grants directly to qualified SCD support agencies for the purpose of increasing SCD patient and family access to psychosocial assistance.

SCD Need: Financial Assistance

The worsening economic climate has taken a drastic toll on national, individual and family finances. The impact of higher food, housing, gasoline, child care and medical treatment costs are obviously more pronounced for those with little to no income. The result is that the need for financial assistance within our society appears to be greater than it has been for many decades. For people with SCD, the attempt to successfully negotiate through the current economic climate may be as difficult as dealing with the disease itself. They clearly need help. In a 1999 paper published in the “Journal of Black Psychology,” Oscar Barbarin, Charles Whitten, Sandy Bond and Rhonda Conner-Warren write, “Financial hardship plays an unmistakable role in accounting for important psychological, academic, and social outcomes often attributed to SCD.” 28

The authors also state, “The effects of psychosocial risks associated with illness severity and poverty are cumulative. Financial hardship and frequent pain episodes combine to produce the highest levels of impairment across all domains. Thus, the poorest levels of disease adjustment and psychosocial functioning occurred for children and parents who experienced financial hardship and frequent pain episodes. These results suggest that the risks associated with poverty and illness are cumulative. The greater the financial hardship and the more frequent the pain, the greater the risk of psychosocial dysfunction.” 29 A great deal of the need for financial assistance is related to the fact that SCD patients and their families, much like many families who contend with other chronic illnesses, have a significant amount of difficulty securing employment. A paper written by Lauren A. Smith and Paul H. Wise published in 2002 by the American Journal of Public Health states, “Welfare recipients and applicants with chronically ill children face substantial barriers to employment related to their children’s illnesses. These barriers include high rates of child health care use and missed work.” 30 The authors also cite another disturbing fact that is a consequence of the inability of parents to receive time off from work. They said, “The economic consequences of work absences could include lost wages or, if absences occur frequently, even a lost job. On the other hand, when parents miss their children’s medical appointments, continuity and quality of care are undermined.” 31
That statement clearly portrays the double-edged sword that continuously threatens the life quality of those with SCD and other chronic diseases. Parents of SCD children often find themselves in the unenviable position of having to choose between going to work or caring for their children. Adults with SCD face another challenge, that of being able to find work with employers that are sympathetic to their conditions and who will allow them flexibility in their work schedules to accommodate the erratic and disruptive behavior of the disease. In these difficult economic times especially, more and more employers are much more concerned with the bottom line and less concerned with the needs of their employees. Even in spite of the anti-discrimination laws and other legislated safeguards for equal opportunity, the reality is that employers value employees who produce and have little or no use for those who do not.

Lyn Ray’s research findings in her article, “The Cognitive and Academic Impact Of Sickle Cell Disease,” succinctly state the importance of this issue. She states, “The more society operates under a model of scarcity and competition, the more parents and their children with chronic illness and disability will have to struggle for access, acceptance, and the opportunity to participate in social and economic life.” Of course, what is true for families with SCD children is probably truer for the adults who cope with it, as well.

There are many other factors that point to the need for society to devise strategies to address the complex financial needs of those who have SCD in their lives. Among these elements is the simple reality that a large percentage of those with SCD possess the ability to generate income on their own and merely need a means to do so. We must do more to help them do that. Conventional forms of employment may not address this need but, then again, SCD is not a conventional disease.

**Solution:** Establish and sufficiently fund a National Sickle Cell Disease Emergency Financial Relief Fund within the United States department of Health and Human Services that will provide federal grants directly to qualified SCD support agencies for the purpose of providing increased financial assistance to SCD patients and their families.

**SCD Need: Occupational Assistance**

There appears to be little research, particularly in the United States, that has been performed to study the occupational opportunities available to adult persons with SCD. However, a 1996 British Psychological Society Paper written by Kenny Midence and James Elander states, “Finance and work are areas in which some of the most severe problems have been reported, and this is reflected in the high levels of unemployment among samples of adults with SCD.” The authors conclude, “People with SCD should avoid work which is physically strenuous, exposes them to extremes of temperature or low oxygen concentration, or which cannot be interrupted to take fluids.
Many perform highly skilled work, but the majority report great difficulty in finding and keeping suitable jobs, and find that SCD places them at a significant disadvantage in the workplace. Although this information is somewhat dated, very little has changed over the years to alter its veracity.

Those with knowledge of SCD understand the occupational challenges that SCD patients face and they understand that the support network for them is virtually non-existent. This is a tragic reality since many SCD patients are actually capable of active participation in the workforce. Midence and Elander bear this out in their statement, “The type of work which affected individuals choose to undertake or are able to obtain is a critical factor, for although the ability to maintain regular attendance at work may be an important consideration in employing a person with SCD, there is no reason why, in a suitable occupation, individuals with SCD should not be expected to perform as well as anyone during long periods of good health.”

One way of interpreting the findings of Midence and Elander is that, in terms of a community’s economic well-being, the lack of occupational opportunity for people with SCD is more of a negative factor than their health. It is important for society to honestly assess, acknowledge and deal with the reasons why employment opportunities for SCD patients are so difficult to find. As Bediako states, “These data suggest that since the rate of unemployment among adults with SCD is greater than the proportion of individuals who report severe pain or who use health care services a great deal, it is likely that factors other than SCD pain or health care utilization account for the unemployment rate in this population.” This is something that needs to be addressed for the good of society as a whole. Employment opportunities must be opened up and occupational assistance must be put in place to help SCD patients find suitable employment that allows them to become as self-sufficient as possible while providing them the opportunity to actively contribute to the gross domestic product of their communities.

**Solution:** Establish and sufficiently fund a National Sickle Cell Disease Occupational Assistance Program within the United States department of Labor that will provide federal grants directly to qualified SCD support agencies for the purpose of providing occupational assistance to SCD patients and their families.

**SCD Need: Entrepreneurship Training**

In that employment opportunities for the majority of persons with SCD are difficult to obtain, it is incumbent on society to identify other means that allow them to contribute to the economy. One of those means is entrepreneurship. The Organization for Economic Co-operation and Development’s (“OECD”) “Promoting Pro-Poor Growth Private Sector Development” report published in 2006 identified the importance of entrepreneurship in eliminating poverty. Much of what is characterized in the report applies to people with chronic diseases like SCD. Among other important statements, the report says, “There is increasing recognition of an emerging pro-poor private sector development agenda that acknowledges that what matters is the degree to which economic growth provides opportunities for the poor and the extent to which poor men and women can take
advantage of these opportunities. Economic and social development are thus interlinked and should be addressed together.”

Another relevant paper entitled, “Chronic Disease: An Economic Perspective,” written by Marc Suhrcke, Rachel A. Nugent, David Stuckler and Lorenzo Rocco was published in 2006 by the Oxford Health Alliance. The paper provides an important overview of the economic burden of chronic diseases from a global perspective and includes the statement, “Overall, a fair amount of evidence exists to conclude that there are important economic consequences of chronic disease – important for the individual and his/her family but also potentially important for the economy at large.”

Certainly, from a macroeconomic point of view, it is safe to say Suhrcke, et. al., are correct in their assessment that chronic diseases like SCD are “potentially important for the economy.” In fact, their assertion is actually understated. That is why it is important to identify ways to develop entrepreneurial opportunities for SCD patients, particularly in consideration of the fact that traditional employment options are so elusive for them. Generally speaking, the intelligence levels and talents of people with SCD are equivalent to those of people who do not suffer from SCD’s physical burdens. The fact that so many SCD patients are unemployed or underemployed equates to decreased productivity and economic growth potential within their communities. In other words, failure to develop ways that allow those with SCD to contribute to the economy has a negative effect on the economy as a whole. Entrepreneurship is one possible solution to this dilemma.

**Solution:** Establish and sufficiently fund a **National Sickle Cell Disease Entrepreneurship Development Program** within the United States department of Commerce that will provide federal grants directly to qualified SCD support agencies for the purpose of providing entrepreneurship training to SCD patients and their families.

**SCD Need: National SCD Legislation**

Most, if not all, of the major social advances made by humanity have come as the result of legislative mandates. Labor laws that have provided safer working conditions, civil rights laws that have provided for equal rights, environmental laws that have provided for improved living conditions and food and drug laws that have provided us with safer food and medicines are just a few well known examples. The power of legislative mandates cannot be overstated when it comes to taking corrective actions to mitigate society’s ills and shortcomings. SCD is one of society’s ills and it requires corrective action. This action should entail the passage of sweeping legislation that will provide for increased SCD research funding, increased education funding, increased treatment funding and increased funding for supportive services.
According to the National Council of State Legislators, fourteen out of fifty states currently have SCD related laws in place but, in virtually all cases, the financial support of these programs is inadequate. The U.S. government also has SCD legislation in place, including the Sickle Cell Treatment Act of 2003 (“SCTA”) which 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.

The Sickle Cell Disease Association of America (“SCDAA”) is currently advocating for more legislation and is asking Congress to provide $13 million for SCTA. As stated on SCDAA’s website, “While this work is progressing, the reach and scope of current activities suffer significantly from underfunding at the national level. SCDAA and its member organizations have noted many States have not moved to implement expanded Medicaid reimbursement provisions authorized within the SCTA. As a result, outstanding needs still far outpace available services.” SCDAA also states, “Throughout this time, full implementation of SCTA programs has received broad based community and bipartisan legislative support. Unfortunately, despite this great support, the authorized programs have never been fully funded by Congress.”

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 for 5 years the authorization of appropriations for the sickle cell disease prevention and treatment demonstration program.”

SCDAA is also seeking $6.5 million for a Sickle Cell Disease and Newborn Screening Program, $40 million for a National Hemoglobinopathy Registry and $10 million for a Sickle Cell Disease Global Support Initiative. All of these efforts and more are desperately needed in the fight against SCD. To be certain, each of the needs identified in the preceding pages of this paper should be addressed through the introduction and passage of legislation that enables not only programs but also adequate funding to support them.

**Solution:** Pass a National Sickle Cell Disease Supportive Services Act that restores funding to the Sickle Cell Treatment Act of 2003 and provides sufficient funding to establish and maintain the programs proposed herein:

- National Sickle Cell Disease Medical Provider Education Program
- National Sickle Cell Disease Social Services Support Program
- National Sickle Cell Disease Emergency Financial Relief Fund
- National Sickle Cell Disease Occupational Assistance Program
It is recommended that the proposed legislation should be written in such manner that qualified SCD support agencies are able to apply directly to the federal government for the associated grants rather than making them available through state and municipal government agencies. This method would eliminate the overhead associated with state government administration and allow for a greater amount of the appropriations to be distributed where it is needed the most.

**Conclusion**

There is a need for a renewed emphasis on increasing the awareness of SCD. There is a greater need to expand public understanding of the many hardships SCD patients face on a daily basis. The needs of adolescents and adults who now live well beyond the life expectancies that were common just a few decades ago are of paramount concern. Not long ago, the life expectancies of SCD patients were much lower than they are now. The result of these longer life expectancies is that there are now many tens of thousands of people in the United States (and millions worldwide) who are not receiving the support necessary to elevate their quality of life. In addition to not receiving this needed support, they are also not being allowed to contribute to their community’s socio-economic well-being.

The sad state of society’s handling of SCD is global and multidimensional in scope. This situation must be altered because SCD’s painful impact on humanity is serious and complex. It is also correctable. History has proven that when change becomes necessary, the desired outcomes can be achieved through stakeholder commitment, community involvement, collective willpower, and passionate, dedicated effort.

Now is the time for such a response to eliminate the menace of SCD from the human condition. It is now time to ease the burdens of the millions of human beings that, through no fault of their own, must live with its sinister manipulations of their lives. Yes, now is the time to make amends for a century of neglect and the racially-oriented social injustice that has been at the root of that negligence. The appropriate allocation and implementation of resources including legislation, funding, research and supportive services can make a difference in the lives of millions and benefit humanity as a whole. That is why we must act now. We must act to reduce the impact that SCD has on its patients, their families, their loved ones and the whole world, as well. We must act now. It is time to stop the pain.

**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 was married to a Sickle Cell patient for nearly twelve years before her death at the age of 36. He has served in various 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.
End Notes

2 Centers for Disease Control and Prevention website; http://www.cdc.gov/ncbddd/sicklecell/data.html.
3 Indiana State Department of Health website; http://www.in.gov/isdh/18110.htm.
4 National Heart Lung and Blood Institute website; http://nhlbi.nih.gov/resources/docs/raredisrupt01.htm#SCD
5 Centers for Disease Control and Prevention website; http://www.cdc.gov/Features/dsSickleCell_ED_Visits/
8 Statement on Signing the National Sickle Cell Anemia Control Act; Richard Nixon. May 1972.
9 National Institutes of Health website; http://report.nih.gov/rcdc/categories/.
11 “Designing a Comprehensive System Across the Life Span: Connecticut’s State Plan to Address Sickle Cell Disease and Trait;” Carey Consulting, LLC In Collaboration with the Stakeholders Group of the Connecticut Comprehensive Sickle Cell Disease Consortium; December 2007, p. 6.
18 “Predictors of Employment Status among African Americans with Sickle Cell Disease;” Shawn A. Bediako; Journal of Health Care for the Poor and Underserved; Volume 21, Number 4, November 2010, p. 1135.
24 “Report to the Arkansas General Assembly;” Arkansas Legislative Task Force on Sickle Cell Disease, August 2010, p. 7.
38. "Chronic Disease: An Economic Perspective;" Marc Suhrcke, Ph.D., Rachel A. Nugent, Ph.D., David Stuckler and Lorenzo Rocco; Oxford Health Alliance; 2006, p. 27
39. Sickle Cell Disease Association of America website.
40. Sickle Cell Disease Association of America website.