

The Abell Report

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Breaking the “Sickle Cycle”

Medical, educational, and employment crises that afflict thousands of Marylanders with sickle cell disease can be curtailed. So why does the sickle cycle persist? Here's what experts say can be done.

ABELL SALUTES:

Jericho, for teaching re-entering prisoners: to regain faith in society, they must regain faith in themselves.

Michael Elliott, out of prison for more than a year now, knows what it is to be down on your luck and in need of help to get on in life. He says, “Jericho does a lot for all of us. But the biggest thing it does is to teach us to have faith in ourselves!” Faith in one’s self, as he describes it, can be transformative: It’s what changes the Michael Elliotts of the world from drifting and in immediate danger of falling back, to moving on and becoming productive citizens.

Often when prisoners are released to re-enter the world they left behind, they find that there is little or no housing for them, no easily accessible medical care, and virtually no way of fitting back into society. What there is, however, is Jericho—a program operating under the umbrella of the Episcopal Community Services of Maryland—and the promise it holds for this population looking for a second chance.

Although Mr. Elliott is a high school graduate, his admittedly poor work ethic cost him his job, his marriage, and his family—and ultimately put him in cir-

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By Joann Ellison Rodgers

Despite decades of medical and public-health efforts, sickle cell disease (SCD) remains a largely invisible, and vastly misunderstood, lethal genetic blood disorder whose victims are mostly comprised of African-American, urban, poor residents, whose status intersects fatefully with failed efforts to improve the overall health, educational opportunities, and quality of life for thousands of Baltimore’s and Maryland’s most vulnerable citizens. The relentless “sickle cycle”—a term that aptly describes the chronic, unpredictable nature of the excruciating and recurring medical crises that mark the progression of the disease—also describes the cycle of underachievement, joblessness, depression, and educational failure that creates undue burden on patients, their families, and the broader community.

Beyond the physical damage and devastating psycho-social trauma resulting from SCD is the sad irony that these effects are occurring at a time when medical specialists can prevent, delay, and/or mitigate the worst symptoms of this most common inherited blood disorder in the nation. Studies by medical and social scientists demonstrate that ignorance, fear, discrimination, and misunderstanding about SCD are largely to blame, and achingly widespread among the very institutions—schools, colleges, emergency medical personnel, employers, community clinics—that urban, poor, and minority populations need most to support them. “This disease is a prototype exam-

ple of how social, health, and economic issues combine to create an environment where poor quality of care yields poor health outcomes and death, as well as poor outcomes in other spheres of life,” says Cynda Hylton Rushton, Ph.D., a nurse, expert on compassionate care, and bioethicist at Johns Hopkins Berman Institute of Bioethics.

This *Abell Report* gathers information from physicians, social scientists, patients, community organizers, and bioethicists to bring needed attention to the plight of people with SCD from public-health experts, policy makers, and opinion leaders. To a person, those interviewed for the article issued a strong call for action to stop the sickle cycle in Baltimore City and the state.

What’s Going On?

Historically, understanding a particular disease’s course, root causes, and symptoms, coupled with public and medical awareness of treatment options, has been a successful paradigm for easing victims’ suffering. It has surely worked to reduce death and disability from heart attacks and stroke; increase long-term quality survival of those with diabetes and childhood cancer; eradicate polio and smallpox; and reduce the stigma, discrimination, mortality, and morbidity associated with HIV infection and AIDS. Among inherited genetic diseases, the scourge of always-fatal Tay-Sachs disease was wiped out in less than two generations after a vigorous public-health campaign to identify carriers of the complicit gene, and those born

with cystic fibrosis, once universally fatal in early childhood, today not only live well into adulthood, but also live well; have access to comprehensive care near their homes; and bear disease-free children of their own with the aid of genetic screening, counseling, and access to reproductive technologies.

Notably, however, in the case of SCD, the historic paradigm has failed many, if not most, of the thousands of Marylanders—nearly half living in Baltimore—who have it or carry the gene for it, as well as thousands of their counterparts across the nation. A century of research, government programs, and physician and patient education have especially failed teenagers and adults with SCD.

State and local newborn screening programs, pneumonia vaccination, and antibiotics have reduced death rates for infants and preschool-aged children with SCD from 10 percent to less than 1 percent in just 25 years. But these survivors now face recurrent, unpredictable, lifelong, notorious bouts of excruciating pain (as one doctor explains it, “on a scale of 1 to 10, this is 11 and up”); multiple and progressive organ damage; repeat and costly hospitalizations or trips to the emergency room; and vastly shortened life spans (42 years on average for men and 48 years for women). Perhaps even worse is the fact that drugs, coordinated care, and psychosocial support remain underutilized, underprescribed, or unavailable for two-thirds or more of those who need them. In Maryland, there is exactly one adult comprehensive-care center, at The Johns Hopkins Hospital in Baltimore, for the entire state, and no center dedicated solely to adolescents.

Similarly, screening for the causative genes to identify carriers is widely available, but underutilized in a population all

too wary of manipulation and genetic testing of any kind, and unreached by trained counselors. In some parts of the state, there are no services available at all by hematologists who specialize in the sickle cell anemia that is the most common form of SCD.

Further, experts claim that teenagers and young adults with SCD experience high rates of depression, unemployment, and educational failure brought on not only by the chronic deterioration of their bodies, but also by the discrimination they perceive—and actually experience—among doctors, teachers, and employers who question their level of pain and the need for opioids to manage it.

Those most familiar with the sickle cycle intensely dislike the label “cyclers” because health-care professionals and others often use it in a way that suggests patients are “repeat offenders” rather than sick people. But the label aptly applies to both the cyclic nature of the disorder itself, and the vicious cycle of underachievement, joblessness, depression, and return trips to hospitals. (By some estimates, the repeat hospitalizations and other care amount to an average of \$40,000 per year per patient. In Maryland, 80 percent of patients live in families who qualify for medical assistance.)

“This is a horrible, dreadful disease afflicting mostly resilient and brave people who the world treats very badly,” says Dr. Sophie Lanzkron, a Johns Hopkins University School of Medicine hematologist and director of the sole adult SCD infusion center and day clinic in the state. (There is also one at Howard University in Washington, D.C.) “In emergency rooms, especially, patients are treated like slackers, suspected of drug abuse, and viewed as criminal drug seekers during devastating pain crises.”

Limited educational opportunities, complicated by poverty as well as pain and

disability, often force those afflicted with SCD into physically stressful jobs like construction and heavy labor, which tax their already-fragile circulatory systems, Lanzkron adds. “I am always filling out forms to get my patients flexible accommodations at work and school. They get fired because their disease causes them to miss so much work. I want to put a protective shield around all of them, and if I sound frustrated, well I am, and so are they.”

Not surprisingly, the physicians, researchers, social scientists, community SCD activists, bioethicists, and patients interviewed for this *Abell Report*, all called for a fresh, urgent look at the sickle cycle by funders, policy makers, medical professionals, voluntary health organizations, schools, universities, and employers. Their view is that the plight of those with SCD needs a lot more attention than it is getting.

The Scope and Impact of SCD

There is virtually no cure for SCD, which at its root affects the ability of hemoglobin-carrying red blood cells to ferry oxygen to every tissue and organ. The abnormal molecules produced by the faulty genetic instructions make fibers inside red blood cells, deforming them from the donut-shaped, flexible discs that easily pass through narrow blood vessels, into crescent shaped, “sickled,” rigid, and fragile cells that break easily. The result of this destruction is decreased oxygenation and cell destruction. Waste products from the detritus of the dying or stricken cells stick around and clog up blood vessels, creating the hallmark episodes of intense pain. During these vaso-occlusive crises, if oxygen is blocked to the brain, then strokes or mini-strokes occur, leading to brain damage, learning disabilities, and other complications. Oxygen starvation injures lungs, kidneys, livers, bones, eyes, and skin, and leads to enlarged and dam-

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aged hearts. Many patients have bouts of acute chest syndrome, in which simultaneous pain crisis in the heart and lungs mimics the crushing pain of a heart attack.

The genetics of SCD are complex because there are several versions of the genes involved that express themselves in a variety of symptoms and degrees of severity. SCD is an autosomal recessive gene disorder, meaning that the disease occurs when two copies of the gene that causes errors in the instructions for making normal adult hemoglobin are present. A person with sickle cell anemia (SCA), for example, the most classical form of SCD, inherits at the time of conception two sets of incorrect instructions—one from each parent—for making a form of hemoglobin, hemoglobin S. Those who inherit just a single copy of this gene, from either parent, are generally (but as we'll see, not entirely) healthy, but become "carriers" with sickle cell trait. A carrier has a one in four chance with each pregnancy of passing on a healthy copy of his or her gene, or an abnormal copy of the gene that causes SCD. Thus, a man and a woman who both have sickle cell trait (each carries an SCD and normal hemoglobin S gene) will have a one in four chance of having a child who inherits only their normal genes, and a one in four chance of having a child with SCD. A one in two chance exists that each child they have will get a normal gene from one parent and an abnormal gene from the other, and therefore be a carrier and have sickle cell trait.

The fallout is not only an unpredictable course of progressive organ damage and pain crises, but also high susceptibility to infections and insidious effects even when there is no pain. The "silent" mini-strokes are particularly destructive in children. "One day," says Lanzkron, "they are great students and then suddenly they have cognitive delays and can't do their academic work because of brain damage from these strokes." Too often, doctors, parents, teachers, and schools "don't get it," she says, "and students are berated for being lazy."

Although the abnormal genes that cre-

ate SCD are present in all socioeconomic groups and races, they persisted especially in populations living in tropical regions of the globe, probably because they conferred some protection from malarial infections, whose parasitic perpetrators infect red blood cells and seem to survive less well in sickle-shaped corpuscles. As forced and voluntary migrations took people from these areas to the U.S., the malaria survival genes came along, together with the likelihood of pairing with another person with one copy. Today, the vast majority of people in the U.S. with SCD are African-Americans clustered among urban inner-city poor communities.

It's not as if SCD is some newly discovered or unrecognized public-health threat. It's been more than a century (1910) since the characteristic "sickled" shape of red blood cells and the painful, damaging anemias and organ damage they caused were identified in a Chicago dental student from Granada, and it's been decades since the populations at risk in the U.S. became clear. (They are mostly, but not all, African-Americans of Sub-Saharan descent, along with Hispanic-Americans and people whose families come from South or Central America, the Caribbean, Turkey, Greece, Italy, India, and Saudi Arabia.)

The late Nobel Laureate Linus Pauling and his colleagues in 1949 were the first to link SCD to an abnormality in the oxygen-carrying, iron-rich hemoglobin molecule in red blood cells, making the disorder the first genetic disease ever linked to a specific genetic mutation. President Richard Nixon signed the Sickle Cell Anemia Act in 1972, pledging to "reverse the record of neglect on this dread disease." In 2003, President George W. Bush signed the Sickle Cell Treatment Act to keep some federal funds flowing for care and research. In Maryland, a 2008 report by the Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease, and a 2006 Department of Health and Mental Hygiene Legislative Report on Adult Sickle Cell Disease in Maryland, included strong calls to action to assist patients and families.

Yet today, for all that history, neither

the state nor Baltimore City—nor the federal government, nor the Sickle Cell Disease Association of America (SCDAA)—knows precisely how many people, especially young and older adults, are living with SCD, owing to the lack of a comprehensive registry already common for other disease categories. Best estimates are that nearly 1,800 adults and 1,700 children in Maryland, with the majority in Baltimore City, are affected. Nationwide, an estimated 80,000 to 100,000 Americans are thought to have SCD—that's one in every 500 African-American live births. Sickle cell trait is believed to occur in an estimated 8 percent of all African-Americans, or two million nationwide. The total economic costs of SCD are thought to be in the range of \$500 million or higher.

The lack of access to comprehensive medical and social services for young and older adults who lack private insurance is compounded by the scarcity of specialists able or willing to care for them. Infants and young children can get comprehensive care at Sinai, D.C. Children's, Johns Hopkins, and University of Maryland hospitals for the first five years of life, but as they age, they "drop not through cracks but a gaping hole," Lanzkron says. Adult hematologists, or blood-disorder specialists, for one thing, generally prefer to care for people with blood cancers—demand is much higher for these services. Several clinicians have been frank in saying the care of SCD patients is "frustrating" and "unrewarding," both financially and medically, given the continuous "crises," emergency room trips, complications, need for counseling, concerns over use of potentially toxic anticancer drugs such as hydroxyurea to treat the disease, and worries (albeit often mistaken or exaggerated) about narcotics abuse. "Adult SCD patients are considered a difficult population among adult hematologists, neither gratifying nor lucrative," says Dr. Suzie Noronha, a pediatric hematologist at the University of Maryland Medical Center and a specialist in SCD care.

For their part, primary-care physicians in the community—both black and white—may avoid treating SCD patients

because the doctors lack expertise in caring for a complex blood disorder, and because the need for frequent monitoring and counseling is a disincentive to treat patients on medical assistance. Noted the 2008 report of The Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease: “The lack of comprehensive medical and social services for adults compromises their health and quality of life...[and] leads to worse health outcomes and increased health-care costs, the majority of which [are] financed by taxpayers.”

As a result, pediatric hematologists are increasingly keeping patients until they are 21 or even older, taxing their already strained resources. “It just breaks my heart,” says Dr. George Dover, chief of pediatrics at Johns Hopkins and a hematologist who pioneered the use of hydroxyurea in children with SCD, “to see these patients get scattered all over, with very few getting organized clinical care or able to participate in research that could help them.”

Finally, the “gaping hole” Lanzkron described has been widening in recent years due to increasing awareness that sickle cell trait is not the benign condition it was long thought to be. Because the afflicted population is expanding, more people are falling through this ever-enlarging hole.

A report in the *American Journal of Medicine* in 2009 by a group of scientists affiliated with Yale University, reported that sickle cell trait occasionally can be linked to “significant morbidity and mortality,” and associated with rare but often fatal kidney cancers. A lack of oxygen to organs, along with exercise-induced muscle damage, releases proteins into the circulatory system that can damage a wide variety of organs. “[C]urrent cumulative evidence link[s] it with damage to the urinary tract, the spleen, fetal loss, neonatal deaths, pre-eclampsia [a dangerous condition of pregnancy], and most of all “exertional sudden death” among amateur and professional athletes and serious exercisers.

The risk of sudden death during exercise is 10 percent to 30 percent higher among those with trait than those without it.

Indeed, the NCAA made testing mandatory for Division 1 athletes in 2010, four years after Dale Lloyd II, a 19-year-old freshman at Rice University died after a football practice, his death due to acute exertional rhabdomyolysis associated with sickle cell trait. In 2010, Benny Abram of the University of Mississippi became the 21st NCAA football player to die of nontraumatic events since 2000, and sickle cell trait has been the leading killer of Division 1 players since that year.

Those with trait are rarely aware of the risks they face, and most are unknown to the medical profession because although screening for trait is conducted on newborns, only those with SCD are notified and followed up with in Maryland.

The Cycle of Disdain and Disrespect

A large part of the sickle-cycle misery derives from the high doses of narcotics required to ease the pain crises, and the medical profession’s scientific and technical inability to verify the intensity of pain and need for narcotics.

Bioethicists who have studied SCD, along with many clinicians, say patients suffer daunting disrespect and denial of pain management, owing to mutual distrust between caregivers and patients that has less to do with education than with attitudes, and racial and class bias. SCD patients’ history of frequent unemployment, and spotty work and school records, feed the bias. Chronic stigmatization leads many to avoid seeking treatment at all.

Rushton, the Johns Hopkins bioethicist, is a member of a research team documenting and looking for ways to address the “cycle of disrespect” she believes “comes from assumptions we make about people based on how they present themselves.” The experience of LaShanta Whisenton, a young-adult patient described in a *Johns Hopkins Magazine* article in 2008, is a typical case in point. “When the pain got too bad [Whisenton] would do what’s done by tens of thou-

sands of people who suffer from sickle cell anemia,” the article noted. “She would gather her medical cards and drive to her local emergency room. But before leaving her home in suburban Washington, D.C., she always did one more thing. She worked through debilitating pain to put on a fresh skirt, jacket, and pair of heels for the trip.... Though she would face an excruciating six or seven hours of waiting before receiving narcotics to numb the pain that sickle cell anemia patients regularly endure, she wanted to appear ‘professional.’” Otherwise, the software analyst and mother of three feared she would be seen as a drug abuser and not get the treatment she needed.

This part of the sickle cycle, Rushton says, begins with “negative provider attitudes about poor minorities and drug abuse that ignite aggressiveness and disrespect in patients themselves, then encourage clinicians to give it back in kind.” The result is a “cycle of broken trust that prevents doctor and patient relationships from being established, and creates discouraging behaviors on both sides. Clinicians fail to see patients as individuals in need, and patients fail to trust the care they are offered,” Rushton says. The same cycle of disrespect also exists outside of medical institutions—in businesses, schools, and social settings.

Gail Geller, Sc.D., also a faculty member at the Johns Hopkins Berman Institute of Bioethics, and a professor in the Hopkins Department of Pediatrics who studies physician and patient attitudes that surround SCD, describes the sickle cycle as “full of Catch 22 moments and unintended consequences.” The level of ignorance among doctors and nurses is so high, says Geller, “I sometimes want to strangle them. There is terrible injustice.” Geller says it would be great to believe that more education alone would help solve the problem, as it has in some ways with other diseases that affect poor African-Americans disproportionately. “But there is something about SCD in which background racism and discrimination intersect more perversely with the disease, possibly because you can’t see or easily measure its

effects, and there are no objective criteria for evaluating pain crises.”

She says research shows that some of the very things SCD patients are taught to do to distract themselves from pain and to cope—listening to music, for example—are misperceived by doctors, teachers, and employers as ‘evidence’ of slacking, inattention, rudeness, or exaggeration of their pain. “When a doctor in an emergency room, or a supervisor at work, or a teacher in the classroom sees someone listening to music, the assumption is that the listener is being disrespectful or feels better than they say they are,” Geller says.

A recent report by Carlton Haywood, Ph.D., a Bioethics Institute social scientist, and an SCD patient himself; and Mary Catherine Beach, M.D., a physician and public-health specialist, and Institute faculty member, described a range of potential barriers to SCD care protocols prescribed by the National Heart, Lung, and Blood Institute of the National Institutes of Health.

According to Haywood and Beach, the list is topped by mutual medical staff and patient distrust and disrespect, fear that a patient is really a drug abuser, a reluctance to prescribe opioids out of fear of scrutiny by law enforcement and regulators, and disbelief in a patient’s self reports of how bad the pain really is.

The study showed the distrust is so bad that in emergency rooms, it’s not uncommon for SCD patients to abruptly or angrily leave, not because of long waits, but frequently, they say, because of the disbelief they face. The dysfunctional distrust also has created what researchers call “pseudo-addiction syndrome.” Triggered by inadequate pain relief, patients feel abandoned and isolated, which in turn leads to acting-out behaviors and anger. For their part, health-care workers become frustrated at not controlling the patient’s complaints of pain, and fearful about inducing addiction and dependence. Over time, the caregivers avoid contact with “sicklers” and “cyclers” as a way to reduce the perceived conflict, and the interloop-

ing cycles of mistrust interact until they reach dysfunctional levels. “SCD patients and medical personnel are two groups in a power struggle all the time,” says Rushton, “and if either one starts exerting more effort or influence, it intensifies the power struggle.” The point, she says, is that both groups have to change. “If we only work on patient empowerment and not on clinician attitudes and practices, we will not get where we want to go.”

Supportive of Rushton’s view about the need to reform physician education about SCD is a 2008 report by the American Society of Hematology documenting that although available guidelines for treating SCD recommend substantial doses of opioids as safe and effective for pain due to vaso-occlusive or sickling crises, “only four of 19 medical textbooks present treatment regimens consistent with those guidelines. Only seven texts note that addiction is infrequent among SCD patients, while 11 of 12 reassure medical students that cancer patients can be trusted to accurately report their pain-relief needs.” In addition, only two textbooks completely define the benefits and impact of hydroxyurea, the first drug approved by the U.S. Food and Drug Administration to treat SCD, and one that increases the amount of fetal hemoglobin. It has been found in repeated studies to reduce sickling crises by half when given regularly to adults. The authors conclude that “most medical texts provide neither adequate information for the treatment or prevention of pain due to vaso-occlusive crises in SCD, nor reassurance of the unlikelihood of addiction in this population.... Misconceptions and prejudices about drug abuse and addiction contribute to this educational void.”

Medical Treatments and Barriers to Their Use

Treatment of SCD, like its symptoms, is layered and complex, owing to the variability and complexity of its several forms and to the relative scarcity of hematologists in private practice who are able and willing to take on the care of adolescents and adults with the disease.

The numerous genetic subtypes of SCD account for varying amounts of normal hemoglobin and the variability of symptoms—differences often poorly understood by families and physicians who don’t specialize in SCD, says Noronha. “When a baby is diagnosed with SCD, it is impossible to know if he or she will develop complications early or late, whether the pain crises will be severe or modest, whether a drug will delay complications or not,” Noronha notes. “Those with some normal hemoglobin-making capacity tend to do better than those with none or less, but even those who have very little often do fairly well, while others in that category are in the hospital every month.”

Blood transfusions are a major part of the experience of SCD patients throughout their lives, commonly used to treat progressively worsening anemia and complications from infections such as enlarged spleens that can burst if not addressed. Some SCD patients need regular transfusions not just to feel better and stronger, but also to prevent fatal strokes and hospitalizations. Frequent blood transfusions, however, pose risks of allergic reactions; contaminations with infectious viruses such as hepatitis and HIV; and iron buildup in the blood, which must be removed to prevent liver damage, heart disease, and diabetes.

Bone marrow stem cell transplants can offer a cure, but only to a tiny number of those who have SCD, in part because there are relatively fewer African-Americans and other minority populations from which to draw closely matched donors (usually relatives) than is the case with diseases that afflict the population at large. It is estimated that less than 1 percent of SCD patients have a suitable matching donor, usually a brother or sister whose immune system’s white blood cells share the same antigens, or proteins. Also, because minority groups are historically more distrustful of the health-care system, they are less likely to volunteer to be donors, and the high cost of transplantation—in the hundreds of thousands—is out of reach for most SCD patients, who

lack insurance.

Recently, scientists have been experimenting with ways to more safely suppress the immune system to allow transplants with “haplo” or half-matched donors, and to find more efficient ways of collecting bone marrow stem cells from babies’ umbilical cords to “bank” and use later. (Stem cells can develop into any cell type, so they can potentially become red blood cells to replenish or can add to an SCD patient’s supply of normal hemoglobin-carrying cells.) Gene therapists are looking for ways to turn off hemoglobin S, or turn on other genes that reset normal red-cell production, or safely implant a normal copy of the hemoglobin gene into bone marrow stem cells to cure the disease in childhood long before organ damage occurs.

But these strategies are far from being widely available. New medicines that increase fetal hemoglobin production are being studied, and drugs such as sildenafil (Viagra), combined with nitric oxide (a chemical needed to maintain blood-vessel tone), have shown a bit of promise in SCD-related lung problems. Other avenues of investigation include nutritional enrichment to address deficiencies of folate, vitamin B6, zinc, and omega-3 fatty acids—common in SCD—and cholesterol-lowering statins that make cells less sticky.

But for most SCD patients, over-the-counter pain relievers, heating pads, fluids, and rest help with mild pain and are the mainstays of their self-care, punctuated by trips to ERs and hospitals for severe pain crises. Standardized protocols, published by the Mid-Atlantic Sickle Cell Disease Consortium (MASCC), part of the Mid-Atlantic Regional Human Genetics Network, call for oxygen therapy, intravenous and oral fluids, and, of course, opioids. However, specialists say that, by far, the treatment that holds the most promise for those with the severest cases of SCD is the one that is most vastly underused: hydroxyurea. The medicine is an anticancer drug that was discovered decades ago to have the interesting “side effect” of prompting the production of

fetal hemoglobin—the kind newborns have—and which is proficient at preventing red blood cells from sickling and causing SCA.

With carefully determined daily dosages, hydroxyurea, which is currently the only treatment specifically approved by the FDA for SCD, has been shown in numerous studies to reduce sickling crises and acute chest syndrome by half, and to vastly decrease the need for blood transfusions and hospitalizations. Recent studies suggest that the drug is also safe and effective in children, and may help them grow normally and avoid or greatly delay organ damage, although studies are still ongoing to make sure this is the case with long-term use.

Unfortunately, hydroxyurea is feared by many patients and doctors. Primary-care physicians often discourage its use because it takes time and skill to monitor its effects, and to recalculate dosages as patients grow and mature. Patients worry that hydroxyurea may cause leukemia or other cancers (which is sometimes the case when used in higher dosages in cancer patients), although research shows that it is safe when dosages are fine-tuned and patients are carefully followed. There is some evidence that hydroxyurea impairs sperm production and fertility, and it also can lower counts of white blood cells, which fight infections.

Experts say, however, that with careful dosing and monitoring, serious side effects can be muted or avoided altogether. Still, they estimate that at least half of those who could benefit from hydroxyurea do not take it and are not given the option by their doctors. In a study published in the *American Journal of Hematology* in January 2011, a team of researchers that included Lanzkron; Haywood; Beach; and Shawn Bediako, Ph.D., a social psychologist at the University of Maryland, Baltimore County, surveyed the attitudes of 94 SCD patients toward the use of hydroxyurea. They found that 70 percent of current users reported substantial or partial improvement, and 80 percent said there were few or no troubling side effects. Of those who took it for

less than six months, 57 percent said they stopped on a doctor’s recommendation or because they did not like the way they felt. The most damning statistic, however, was that 50 percent of those who never used hydroxyurea said they had never been given information about its benefits, and 85 percent reported they believed they would get no relief from taking it.

At Lanzkron’s adult clinic, 70 percent are taking hydroxyurea, thanks to the rigorous counseling of patients, but she acknowledges that it is sometimes a struggle to get patients to try it. Efa Ahmed-Williams, a knowledgeable community SCD activist and director of Destiny Despite Sickle Cell Program in Baltimore, is a reluctant user at age 35. “I finally agreed, but it was not an easy decision because of dosing issues and the need for lots of monitoring,” she says.

The Pain

The “holy grail” of SCD, says Lanzkron, is finding a standardized, sure-fire way to measure the intensity of SCD pain. So far, she says, “no such test is on the horizon.” Lacking such a tool, those who treat SCD pain rely on patients’ descriptions, which are not pretty. One 38-year-old person afflicted with the disease, quoted in a *New York Times* article in the summer of 2011, likened it to “a jackhammer on your back throughout your whole body... [s]ometimes...in your joints, in the abdominal area, in your head, in your chest.” Latoi Walker, a 35-year-old Baltimorean, called it “the kind of pain you wouldn’t wish on your worst enemy,” a “deep, bony pain” relieved only by the strongest opioids given intravenously over many hours. Carlton Haywood, the Hopkins bioethicist, walks stiffly because of damage to his joints and hips. He describes the pain as “the worst ... you’ve ever had ... beating all over your body like a steady... rain, with lightening flashes.” One of Lanzkron’s patients told her that the multiple broken bones he suffered in a 20-foot fall on his construction job “couldn’t come close” to the pain of a sickling crisis, and another says it is “like having nails driven into your joints.”

Moreover, the pain of SCD is not an “on-off” phenomenon in which periods of pain are separated by longer periods without any. “SCD patients are always managing pain or worrying about managing pain,” notes Geller, the Hopkins researcher who helped create a training video and curriculum designed to sensitize health-care workers to the sickle cycle, and who has spent hundreds of hours with SCD patients.

Long waits for care in ERs further aggravate a sickle crisis because cells undergoing acute spikes of oxygen starvation begin to die, adding to the cumulative damage and causing higher risks of strokes, infections, leg ulcers that can require skin grafts, gallbladder disease, blindness, and bone damage. The revolving-door nature of SCD pain therapy causes post-traumatic stress disorder in some, and psychological and behavioral problems in others.

Further complicating the pain problem is the variability from episode to episode in a given patient and among patients, a phenomenon that adds to the confusion and mistrust among caregivers. Triggers of pain episodes are unpredictable and also variable. According to Lanzkron, in one group of patients, exertion, heat, and cold could trigger crises; in another group, these variables could have no effect. “Many cannot find the words to adequately explain what is happening to them, so they just stay silent,” she says.

Not only is the pain caused by SCD a challenge, but the medicines that are used to manage the pain—Dilaudid, morphine, oxycodone, and hydroxyurea—can be problematic as well. Ahmed-Williams, the SCD activist in Baltimore, requires pain management at Lanzkron’s clinic about once a month, and has logged a lot of “sick days” because of the side effects of medications. The well-educated wife and mother of a 6-year-old was diagnosed with SCD at age 2, and says she was told she would probably die before her 20th birthday. “I beat the odds every day,” she says, “but there is a price.”

Education and Employment Challenges

Even when economic indicators are good, education and employment figures among those with SCD are not, and in times of economic downturns, obstacles to financial security and higher education opportunities can seem insurmountable to adults with the disease. “The big, big problem for SCD patients is getting a job, keeping a job, and advancing in a job,” says Bediako, the University of Maryland social psychologist who has devoted much of his career to documenting and studying barriers to employment and health-care services among adult SCD patients. Along with Carlton Haywood, he serves on the Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease; Bediako co-chairs the committee.

In focus groups and surveys conducted by Bediako throughout Maryland and elsewhere, he and his team have found “inordinately high rates” of unemployment among adults with SCD. Hard data are scarce, but where available, they are scary, he says. “We estimate that Sophie Lanzkron’s clinic patients experience a 45 percent to 60 percent rate of troubled employment or unemployment.” While some would say this is to be expected among people with chronic illness, Bediako and his colleagues say that what’s at play with SCD is often absent in those with other chronic disorders—namely, the unpredictable intermittency and duration of symptoms.

For example, Bediako says, a pain crisis may occur on a Tuesday, and although the worst may be over on Wednesday, it’s still not entirely gone—plus, the Monday before the crisis may have been disabling as well. “For many with SCD, the precipitating symptoms that build up to a crisis can hold them back from work or school in an effort to mitigate or stop a full-blown crisis,” he says. “If the crisis occurs anyway, time is needed to bring the crisis down, often a day in the infusion center or an emergency room or hospital bed, and then there is a recovery period. The bottom line is that SCD requires exceptional levels of trust and flexibility

between and among patients, schools, and especially employers who may be more comfortable with a predictable pattern of sick leave or employment.”

Noting that neurobiologists have shown that stress exacerbates physical disease, he says, “the sickle cycle is mostly meant to describe pain and stress, but I see it more comprehensively as pain, stress, and barriers to a good education, and to finding and keeping a job.”

Bediako is especially concerned about the ignorance among employers, and among SCD families regarding patients’ rights under provisions of the Americans with Disabilities Act (ADA). Many, he says, who have their hours cut or are fired may have recourse in law and regulations if there have been no reasonable accommodations made for their disease. “But if you are not seen in a comprehensive clinic that has social workers, you won’t likely get information about ADA.” The goal is not more lawsuits, he says, but education. “If more efforts were made to accommodate people with SCD, we would see a big shift in the financial burden that SCD places on the tax system of Medicaid,” Bediako says, “because when people are employed, they get access generally to better insurance and better care that can reduce absences and decrease the need for accommodations from employers.”

Bediako has conducted studies to document other factors that affect unemployment in SCD patients. In the November 2010 issue of the *Journal of Health Care for the Poor and Underserved*, he analyzed “predictors” of unemployment gleaned from a survey of 115 adults with SCD who are part of a long-term cooperative study of SCD. The analysis showed that women were almost three times more likely to be employed than men, and the odds of being employed increased by a factor of 1.47 for each one unit decrease in assertiveness. He concluded that certain elements of modesty and timidity (just the opposite of the behaviors one often needs to get adequate pain care or push through appropriate requests for accommodation or days off) are more likely to get and keep SCD patients employed.

“If I could only pick one area in which to intervene and make the biggest impact for young adults, I would start with schools,” Bediako says. “If we start early to build positive educational expectations and provide support for students with cognitive problems and pain, we will also go a long way to reduce their overall health and economic status.”

Carlton Haywood is exhibit “A” for Bediako’s point of view, having had teachers, he says, who expected much of him despite his diagnosis, and—with some pushing from his doctors and parents—showed flexibility regarding his attendance. “I know I am more fortunate than many working adults with SCD,” writes Haywood in a recent Web essay for the Bioethics Institute. “At a personal level, my colleagues completely understand and empathize when symptoms sideline me.” He adds: “At times, health researchers, me included, pay too little attention to anything beyond what happens when patients seek medical treatment...I can attest that for my fellow patients and I, medical treatment can at times be the least of our worries...[A] broader approach to exploring the needs of patients, one based on social justice concerns, is needed, whether they have sickle cell disease or a different chronic...condition.”

The Durable Legacy of Mistrust

According to a 2010 article in the *Journal of Health Care for the Poor and Underserved*, compared to the U.S. population in general, people with SCD are almost three times more likely to say they are shut out of decisions involving their care. Some 53 percent say they are not always treated with respect, compared to 34 percent of the general population. Further, 58 percent say they don’t get enough pain relief, and 72 percent say it is hard to get help when they need it, compared to 17 percent and 37 percent, respectively, of the general population.

Racial and class discrimination certainly play a role in such experiences, but it is difficult to tease out the impact that can be

shown for other health-care delivery disparities, such as those found in diabetes or cardiac care. One reason: There are no “majority” populations against which to compare the largely African-American minority affected by SCD. The point here is that there is no “white” or other population with SCD against which to compare the treatment of African-Americans with the disease, as is the case with diabetes, or hypertension, where race disparities in care are clearer because there are both blacks and whites with those disorders.

The mistrust between caregivers and patients is also fed by the history of misguided, albeit well-intentioned, efforts by state, local, and military programs to introduce screening and testing for SCD and sickle cell trait.

Dover, the hematologist at Hopkins who pioneered the use of hydroxyurea in children with SCD, points to a military screening program for trait in the 1970s that led to discrimination against those who had it, including career-busting bans from certain duties and ostracism. When research later showed that the health risks from trait could be eliminated with good hydration and training modifications, the restrictions were dropped, but the damage to trust among African-Americans at risk remained. State agencies, advocacy groups, and community-based organizations “established many voluntary screening programs without appropriate foresight or provisions for education and counseling, and often caused confusion by disseminating misinformation regarding the difference between a diagnosis of sickle cell anemia and the presence of sickle cell trait,” Dover said in a recent article published in the September 2010 issue of *The New England Journal of Medicine*. “These programs were thought to do more harm than good and have been abandoned or heavily modified.”

A newer case in point, according to Dover, is the NCAA’s sickle cell trait screening program, which “poses a cautionary tale” despite the good intentions of its organizers. He says the program, which expected to screen 170,000 college athletes and find 400 to 500 new cases

each year, could—at its best—benefit individual health and help carriers make “informed reproductive decisions,” but “like its predecessors...[is] full of potential pitfalls.”

Among the unanswered and worrisome questions the NCAA program raises, says Dover, are whether false positives will be eliminated by secondary testing; how the knowledge of being a trait carrier will affect student athletes and their families; and whether athletic directors will protect student privacy, or alter training and play schedules to limit or eliminate all or some of the risks. “[T]here is now great interest in genetic-based risk profiling and personalized medicine,” Dover says, “and screening programs such as the program the NCAA has instituted could lead to an epidemic of testing for genetic risks about which nothing can be done or that would lead to discrimination against players not just on the athletic field, but in their ability later to get insurance, or particular kinds of jobs later on.”

Community-based voluntary health organizations have the potential to vastly improve access to screening, counseling, and treatment, but the very act of seeking information and care by the urban poor minorities who make up the bulk of the patient population can be their undoing in schools and in the workplace, according to Karen Proudford, Ph.D., president of the Baltimore-based William E. Proudford Sickle Cell Fund, which is named for her father. She herself has trait. For Proudford, raising awareness in the SCD-prone population is necessary, but it is also part of a double-edged sword not easily blunted.

The mistrust and discriminatory history attached to SCD has also sabotaged efforts to recruit African-American athletes, celebrities, business leaders, and other “champions” who might put a compelling face on the needs of patients, and strengthen government and private financial commitments. The Sickle Cell Disease Association of America does a fair job of leveraging what spokespersons it can find, but the small group of singers, athletes, and actors in that category are not the “household names” that have boosted

other diseases into the national consciousness, such as Jerry Lewis (muscular dystrophy), Michael J. Fox (Parkinson's disease), Boomer Esiason (cystic fibrosis), and Magic Johnson (HIV/AIDS).

Says Proudford: "The cycle of cynicism—among doctors, nurses, teachers, and bosses—persists. This disease silences you, and is especially heartbreaking in young people, who learn not to voice their pain. That is the enduring legacy of mistrust."

A Tale of Two Diseases

SCD is a so-called orphaned disease. It is so relatively rare, and afflicts such limited populations, that it presents little incentive for large drug companies or research enterprises to search for marketable treatments. Such corporate behavior is not difficult to understand. Harder to accept, perhaps, is why SCD has been orphaned at the patient-support and community-advocacy levels as well.

Compared to SCD, other rare, relatively rare, or incurable diseases—such as cystic fibrosis, muscular dystrophy, and Parkinson's disease, for example—have far stronger national, celebrity-studded "public faces;" support from private foundations, philanthropists, and broad grassroots volunteer organizations; and highly publicized events such as telethons and pro-am golf tournaments that draw attention to their cause, and facilitate lobbying for more research and fundraising.

In particular, for those in the SCD community, the starkest contrast is found in the public visibility and support for cystic fibrosis (CF), an even rarer autosomal recessive disorder. CF, like SCD, is inherited at birth and strikes 30,000 Americans—less than one-third the prevalence of SCD. Further, CF occurs in one in 4,000 live births, compared to one in every 500 live births for SCD. Single copy CF gene carriers affect one in 30 Americans—almost all of whom are white—compared to one in eight African-Americans who have trait.

The 30,000 to 35,000 children and adults with CF in the U.S. have access to a

national network of 150 multidisciplinary centers, while three times as many SCD patients have access to just a few, including those at Hopkins and Howard University, and one each in Georgia and North Carolina. Interestingly, a 2009 study by Scott Grosse, published in the journal *Pediatrics*, identified 130 comprehensive-care centers serving just 15,000 hemophiliacs, compared to just 10 for 100,000 SCD patients.

Grosse's findings shed further light on the racial disparity inherent in the contrast between community and medical support for CF and SCD. In the study, he and his team looked at access to comprehensive care for several genetic disorders, and showed that for hemophilia and CF, which largely afflicts whites, "effective national networks of specialty clinics exist and reach large proportions of the target population." For other disorders, however, "notably sickle cell disease, fewer such centers are available, are less likely to be networks, and are used less widely," despite their wider prevalence and the location of affected populations in concentrated urban areas with large numbers of health-care providers.

Those who study social, racial, and economic factors that influence disease outcomes, also say that people with SCD avoid public exposure and often hide their disease status, while CF patients on the whole do neither. Former British Prime Minister Gordon Brown, whose child's CF diagnosis was publicized as part of the hacking activities now scandalizing Rupert Murdoch's tabloid empire, was outraged not at the fact that his child was identified as a CF patient, but rather how the information came to light.

Local SCD community organizations trying to cultivate the kind of attention paid to CF acknowledge that a large part of the problem is the fragmentation of their efforts. In Baltimore and Maryland alone, there are half a dozen separate groups involved in patient outreach, fundraising, and lobbying. These include the Lauren Beck Sickle Cell Foundation in Glen Burnie, the William Proudford Sickle Cell Fund, the Francine Allen-Franze Fund for Adult Sickle Cell Disease

Research, and the Destiny Despite Sickle Cell Program. Each has a separate website, and separate activities and offerings, and none has yet officially affiliated with the SCDA national headquarters in the city.

This fragmentation exists in contrast to the strong, centralized visibility of the CF Foundation (<http://www.cff.org/>), and its carefully protected brand identity and local affiliate chapters. The Sickle Cell Disease Association of America (<http://www.sicklecelldisease.org/>), headquartered in Baltimore on East Baltimore Street, is sometimes alternately referred to on its own website as the National Sickle Cell Disease Association, and is sometimes confused with the American Sickle Cell Anemia Association (<http://www.ascaa.org/>), a United Way organization. SCDA's focus is on nationwide awareness and lobbying on Capitol Hill for increased funding, while other community SCD groups focus on local education and direct services to patients. These organizations' leadership and members interact, but their programs and agendas operate separately.

Sonja Banks recently became president and chief operating officer of the SCDA in 2010. She has 20 years of nonprofit management experience with the United Way and the United Negro College Fund, and says she fully "recognize[s] the need for a stronger, centralized presence to aggregate the SCD community's visibility, clout, and impact."

According to Banks, the SCDA, with about 50 affiliates in the U.S., is "improving," even though the organization has long been beset by financial and management challenges. Its last annual posted report, for 2009, documented financial declines over the past five years, owing in part to the recession, and total assets of \$1.52 million. It described having to close several community member organizations and cut staff, as well as borrow \$115,000 from its \$290,000 investment portfolio, and reported public contributions of \$653,300 nationwide.

Meanwhile, the Cystic Fibrosis Foundation has 75 well-established chapters, 110 CF care centers, 10 CF Foundation-

supported research centers, and 55 affiliate programs. Some 96 of CF's offerings are for adult CF patients. In 2010, the CF Foundation received \$118 million in public contributions and posted net assets of \$188 million, an amount that was flat but not down over 2009 figures.

Banks and her colleagues struggle to explain this disparity of public support, given the higher prevalence of SCD over CF. Perhaps part of it is that the national SCDA, which relocated from California to Baltimore just seven years ago, is reluctant to interfere with established community organizations' fundraising efforts. Chronically cash-strapped, these organizations may avoid joining the national group to save on dues; however, Banks believes that the SCDA's lobbying mission, with increased funds, could be more effective.

"There is no doubt fragmentation of SCD organizations hurts," says Banks. There is a great need, she says, to coordinate efforts to persuade states, including Maryland, "to establish more comprehensive treatment centers, and to do more to unify data gathering and public awareness." Without coordination, she adds, all of the organizations are handicapped in their efforts to promote activities and programs potentially fundable under terms of the Sickle Cell Disease Treatment Act. For example, states can apply through the federal government to receive matching Medicaid funds for drugs and other SCD care. But according to Banks, only about seven states have applied so far, not including Maryland.

While there is a certain amount of envy at the success of the CF community, Banks, Proudford, and Ahmed-Williams emphasize that the goal is not to "pit one disease against another" in a zero-sum game to gather resources for one group of patients at the expense of another. Instead, they aim to learn from the CF Foundation's successes within its community and apply that knowledge to hopefully bring more attention to those battling SCD.

Conclusions and Recommendations

By all accounts, the biggest challenge that remains to bending—if not breaking—the sickle cycle in adults is not necessarily developing new treatments, even though they are needed. Rather, multiple challenges exist and include the following: access to and use of existing treatments; mistrust and hostility between physicians and patients fed by negative stereotypes regarding the use of opioids for pain; the perceived and actual racial and social discrimination against people with SCD; the relative absence of accommodation and flexibility by educational and business organizations for a population already at socioeconomic risk; the perceived and actual insensitivity among caregivers to the plight of people with SCD, which has the effect of sabotaging compliance with treatments, undermining pain-coping skills, interfering with efforts to screen for SCD and trait, and discouraging participation in clinical trials; a relative loss of interest and momentum by local, state, and federal government agencies in supporting SCD initiatives; and the absence of coordinated or unified national- and community-based support and service organizations that can effectively lobby for more resources, research, and attention.

Despite these barriers, those interviewed for this report say there are tools and tactics available to make life considerably better for thousands of people with SCD in Baltimore City and Maryland.

Here are their recommendations:

Provide education about SCD to patients, health-care providers, and the public

- Increase funding to initiate or expand sensitivity training and education of emergency room physicians and nurses, employers, and school officials. One novel tool, developed by a team at Johns Hopkins, is comprised of professionally produced videos of teenage and young-adult SCD patients recounting their everyday lives and experiences at The Johns Hopkins Hospital. Initially

funded by a grant from the Niarchos Foundation, the videos include commentary by hematologists, psychiatrists, and social workers, and there is an accompanying curriculum guide. Five years in the making, the video project was designed with the idea that doctors and nurses "are good people who don't want to do harm," notes Geller, faculty member at the Johns Hopkins Berman Institute of Bioethics. "The goal was not to point fingers and pin blame but to help caregivers reflect privately on their behavior with SCD patients." Preliminary evaluations of the impact of the training on physicians and nurses show changed attitudes and behavior, although more research is underway to determine whether the changes last, and if so, how long. The goal, Geller says, is wider distribution of the program, and creation of collateral curricula for use in schools, colleges, churches, and business settings. "An eight-minute film can't change the world," Geller says, "but we know it can have a real impact."

- Educate patients, along with minority and nonminority physicians, to increase sufficient use of hydroxyurea and opioids. "We need physician education about this, but also public education that encourages patients to ask for, even demand, better use of drugs that can help," says Noronha, a pediatric hematologist at the University of Maryland Medical Center.
- Re-create in-service training for principals, teachers, and school nurses in the care and support of SCD patients. These have been successful in the past, according to Dover, chief of pediatrics at Johns Hopkins, but were pretty much abandoned over time. Ideally, experts say, this is a winning activity for SCD advocacy groups and specialists in SCD care.
- Convene a "game changing" conference that brings together every SCD stakeholder: patients, family members,

church leaders, civil-rights experts, Chamber of Commerce officials, major insurers, HR directors, political leaders, teachers, employers, medical-care providers, public-health officials, medical organizations, hospital officials, and representatives of SCD community and service organizations. Rushton, a bioethicist at Johns Hopkins Berman Institute of Bioethics, says the conference, with the right facilitator, would get everyone talking about the problem, help blend disparate agendas, and organize priorities. “It may seem at first like getting a lot of odd couples to come to marriage counseling,” Rushton says, “but SCD care is a complicated environment and new models are needed. If we don’t do something different, we’ll be having the same conversation 20 years from now.”

- Pay attention to the “Oprah Factor” by coordinating professional and voluntary efforts to recruit highly visible champions to take the SCD story to the public. The wish list focuses on prominent African-Americans, including Will Smith and Jada Pinkett Smith, Denzel Washington, an NBA star, and—as always—Oprah herself. (A Google search for “famous people & SCD” revealed fewer than 10 names, several of whom (jazzman Miles Davis and Temptations singer Paul Williams) are deceased.

Strengthen and reform the delivery of medical services to patients with SCD

- Re-energize state and local government agency plans to increase the number of, and access to, comprehensive-care centers for teenage and adult SCD patients. Lanzkron and others suggest two more adult centers, one in Prince George’s County and one on the Eastern Shore. Her current center, at Johns Hopkins, has an annual budget of about \$1 million and cares for 400 or more adult patients. The keys to success in this realm, experts

say, are partnerships between SCD community-action and care groups, Maryland’s two academic medical centers, hospitals already providing SCD care to newborns and young children, and public-health officials at the state and city level who have opportunities for federal and state grants.

- Establish at least one specialized clinic for adolescents. “Teenagers with SCD too often become lost souls, and we lose track of them,” says Noronha. “Some studies show shortened life expectancies are closely linked to this period of transition to adult care, and adult clinics aren’t good at managing teenagers.” Experts point out that when children reach teen years, there is generally less compliance with care regimens, and teens with SCD, like all teens, don’t want to be different—to give up sports, stay home from the prom, or go to the nurse’s office to take medication. Adolescent SCD patients also have increased anxiety and depression. Johns Hopkins is designing a program for this population, and Noronha says the University of Maryland has a clinical psychologist who works with her teen patients. Opportunities to use PDA technology, text messages, and other social media to communicate with SCD teens are ripe for the picking, she says.
- Focus special attention on SCD patients as they age out of pediatric care and “fall through the cracks” of the health-care system, transitioning through adolescence and into young adulthood. University of Maryland’s Noronha calls these patients “an especially vulnerable population most in need of support in educational and occupational ambitions, health care, reproductive counseling, and emotional support, as well as access to Medicaid or other insurance, home-care assistance, and training in self-management of pain crises.” Both Bediako, a social psychologist also at the University of Maryland, and Noronha say middle

schools and high schools are “ideal places” to locate social-support programs during this transition for middle and high school students.

- Assign a “medical home” for every patient who leaves pediatric care. Lanzkron suggests a “hub and spoke” system comprised of three comprehensive adult centers and a network of community hospital-based and private physicians who become the primary-care doctors for transitioning SCD patients. Specialists say such a system would save money by reducing costly hospitalizations among the uninsured.
- Initiate public-awareness campaigns to recruit patients and families for participation in clinical trials of drugs and other treatments.
- Focus on the goals of the Maryland Statewide Steering Committee on Services for Adults with Sickle Cell Disease. The committee, in its 2008 report, recommended a statewide patient registry and a plan to find a medical home for each patient; ensuring access to Medicaid or low-cost private insurance; development of standardized treatment protocols for emergency departments and urgent-care facilities; and the shifting of fragmented resources toward comprehensive and preventive-care models such as regional infusion centers. Bediako says a new report is being prepared to re-energize the work of the group and implement recommendations.

Provide supportive services to patients with SCD

- Spread the word about the rights of SCD patients under the Americans with Disabilities Act. A partnership between community groups and social workers connected to health-care organizations could initiate public-awareness campaigns to help those with SCD better understand their rights.

- Develop peer-to-peer mentoring programs in schools and community centers. One approach, Noronha says, that has worked well in her pediatric population is to connect families who are reluctant to use hydroxyurea to families whose children are successfully using the drug. “If the children are matched in age and gender, the impact is effective,” Noronha says. She cautions that there are ethical issues to be addressed to avoid pressuring families into taking drugs they don’t want, but similar programs would be useful for older SCD patients as well. “I can see peer-to-peer mentoring at the college-student level, by members of the Chamber of Commerce, and by older SCD patients who are working at a variety of jobs.”
- Support efforts to coordinate and integrate the work of patient-advocacy and community SCD service organizations. One model, says Proudford, president of the sickle cell group named for her father, is the Komen Foundation, which is successful at both fundraising and patient services. In the short term, the various SCD groups in Baltimore and Maryland could, during Sickle Cell Awareness Month each September, coordinate programs and distribute awareness materials at grocery store chains, sports organizations, churches, and inner-city school PTA meetings.

EDITOR’S NOTE:

A copy of the report, *Breaking the “Sickle Cycle”: Medical, educational, and employment crises that afflict thousands of Marylanders with sickle cell disease can be curtailed. So why does the sickle cycle persist? Here’s what experts say can be done*, is available in “Publications” on the Abell website, www.abell.org.

Joann Ellison Rodgers, an award-winning science journalist, directed Johns Hopkins Medicine’s media relations and public affairs division for 25 years, and now serves as senior advisor. A graduate of Boston University and the Columbia University Graduate School of Journalism, she is a board member and past president of the Council for the Advancement of Science Writing; past president of the National Association of Science Writers; a Fellow of the American Association for the Advancement of Science (AAAS); and a member of Sigma Xi, the Scientific Research Society. The author of seven books, including *Sex: A Natural History* (Henry Holt and Company, NY), she has contributed articles on medicine, genetics, risk communications, and psychology for *The New York Times Magazine*, *Psychology Today*, *The Los Angeles Times*, and other magazines. Her awards include a Lasker award for medical journalism.

ABELL SALUTES

continued from page 1

cumstances where his life fell apart. In 2008, he was incarcerated for possession of drugs and was released in March 2010, and through good fortune, found himself in the Jericho family, which is centered at 901 N. Milton Ave. in East Baltimore. What happened next is a case study of Jericho. Having lost faith in the institutions he needed to get on with his life, it was Jericho that taught him to, first, have faith in himself. It is in this context that the work of Jericho, and the life stories of the Michael Elliotts, is best understood.

Jericho is, first of all, a workforce development program designed to assist ex-offenders re-entering the community and willing to participate in a two-week job-training, orientation, and job-placement program. The program serves more than 200 men a year and boasts a job-placement rate of 60 percent. Among the positions in which Jericho has been successful in placing its participants are: roofer, warehouse worker, utility worker, driver, cook and waiter, meat wrapper, chef, housekeeping situations, floor technician, automobile detailer, assembly line worker, telecommunications technician, store clerk, appliance installer, electrician helper, receptionist, and carpenter’s helper. Companies where Jericho participants are working include: Danko Arlington, 2nd Chance, Moveable Feast, Atlantic Lift Truck EPA Healthy Homes/Renovator, Jump Start, CCBC Construction Apprenticeship Training,

Fitch Company, and Floor Tech Training.

However, to enter the Jericho program, to get and hold a job, and to live a productive life, these ex-offenders need housing—and given their circumstances, obtaining housing on their own is nearly impossible. So in partnership with The Abell Foundation, Jericho is able to provide transitional housing to homeless clients, to give them the stability they need to make the most of any newly found employment opportunity.

But it is the personal one-on-one successes—Mr. Elliott, for example—that make up the overall success of Jericho, and these are due in no small measure to Jericho’s ability to inspire clients not only to accept the reality of their situations, but also to learn how to “fit in,” even if it’s their second or third time around. Mr. Elliott explains: “For me, for many if not most of us who graduated from the Jericho two-week program, and got a place to live and a place to work, and an opportunity to live a productive life, the journey is largely spiritual. Before you come to believe in the program’s possibilities, you have to come to believe in yourself. You have to learn to have faith in you.” The hundreds of ex-offenders who re-enter society through the gates of Jericho appear to agree with that view.

Abell Salutes Jericho, and the Episcopal Community Services of Maryland, for teaching re-entering prisoners how to regain their place in society—through the acquisition of housing, steady employment, and ultimately a new life—by regaining faith in themselves.