Facts on Sickle Cell Disease

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Joint Budget Hearing for Health/Medicaid
January 29,2020

NYS and Sickle Cell Disease

For 9 years the sickle cell disease bill has languished with the Governor and the legislature. It is the major health care disparity in NYS which has continued to be ignored. There is minimal state funding specifically for sickle cell disease and has consistently been cut over the last 20 years, from $500,000 to $170,000. This despite the fact that the median age of death for severe sickle cell disease has decreased and in males is 38 and females is 42, the severe morbidity of sickle cell disease which prevents many from completing schooling and obtaining/maintaining jobs, that NYS has 10% of the nation's sickle cell disease population but NYS is the worst in providing funding for sickle cell disease and finally, most sickle cell disease patients are on NYS Medicaid and improving the care could save Medicaid at least $4-$5 million a year. In a year where the Medicaid budget is needing to be trimmed, improving the care of patients with sickle cell disease while decreasing the Medicaid budget should be obvious.

Finally, the Assembly graciously gave a one year increase in the sickle cell budget in April last year. This went to the Dept. of Health to distribute but they did not notify groups of an increase in their funding until Nov. 2019, giving them only 4 months, instead of 12 months, to spend the money allocated. In addition, the senate gave an additional $250,000 in June 2019 to several sickle cell CBOs, but as of the writing of this testimony some of the groups still have not received this money from the Dept. of Health and will have less than 2 months to spend this money. This really is not the ideal way to help sickle cell patients and community organizations. When money is allocated, it should not take 8 or more months of a year of spending for the money to be sent to the organizations.

Approximately 100,000 people live with sickle cell disease (SSD) in the US, with approximately 10% living in NYS.

Sickle cell disease is most common in African-Americans, but also occurs in Hispanic, Mediterranean, Middle Eastern and Indian communities. In NYC Tibetan and Asian communities have also been affected.

Births	United States	New York State
African American	1:365	1:230
Hispanic	1:16,300	1:2,320
Caucasian	1:80,000	1:41,647

In NYS, 1:1,146 live births have sickle cell disease with 12% of those births in the Hispanic community. Higher birth rates occur in mothers who were born outside of the US.

Genet Med. 2013; 15:222–228

Approximately 80% of individuals diagnosed with sickle cell disease in NYS live in the NYC **area.**
76% of newborns were born in NYC and 24% of newborns were born outside of NYC
Genet Med. 2013; 15:222–228
https://www.cdc.gov/ncbddd/sicklecell/documents/SCD_in_NY_Prov.pdf

3,000,000 people in the US have sickle cell trait. Approximately 1:12 African Americans have sickle cell trait.

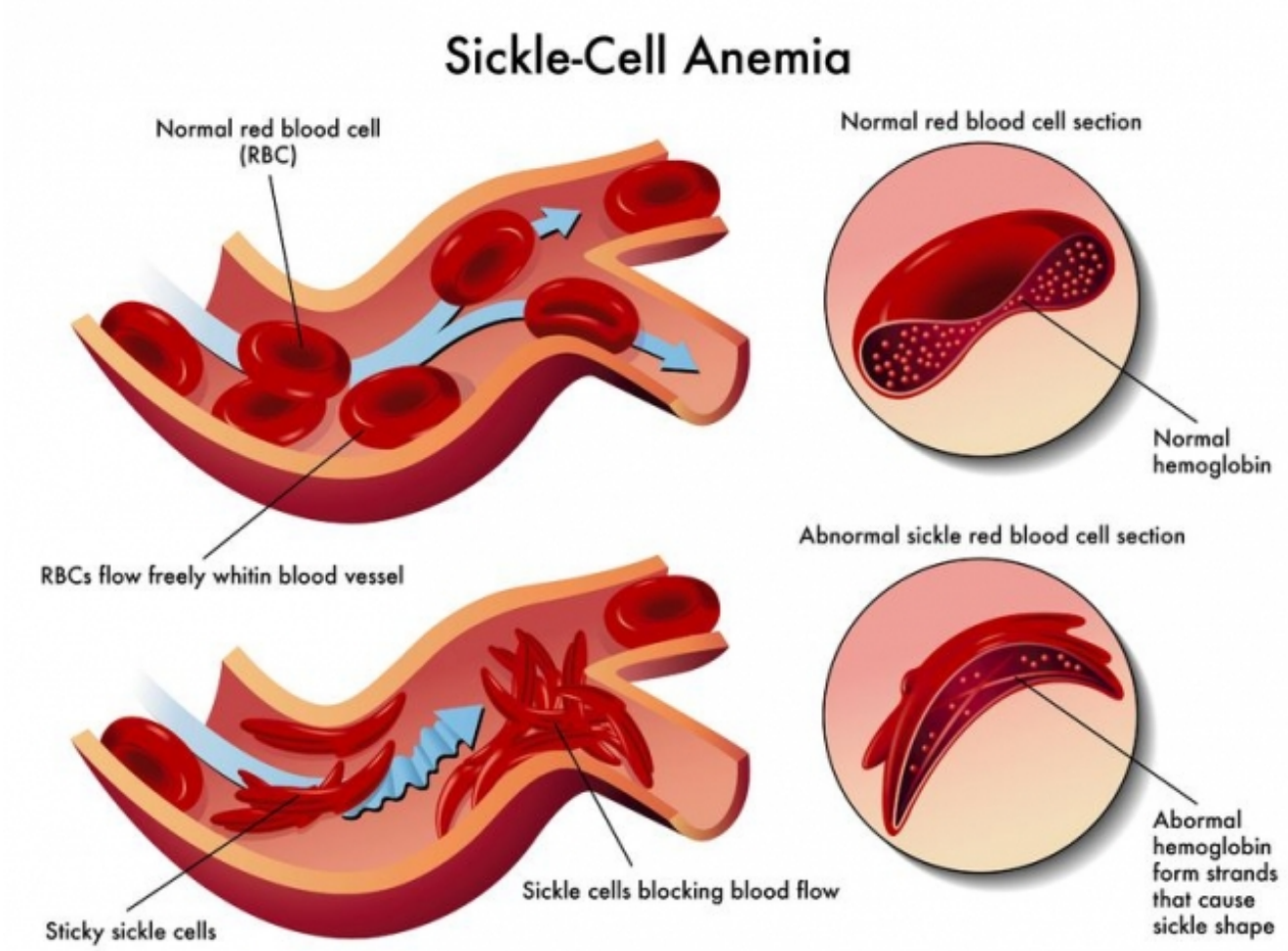
What is Sickle Cell Disease and What are its Complications?

SSD is an inherited blood disorder that is caused by a mutation in the hemoglobin protein (the part of the red blood cell that carries oxygen throughout the body). There are 4 main types of sickle cell disease – SS, SC, $S\beta^{+}$ Thal, and $S\beta^{0}$Thal. To have a child with sickle cell disease one parent must have the sickle gene (S) and one must have one of the other genes (S,C, βThal). A person who has one sickle gene (S) and one normal gene (A) has sickle cell trait (AS).

The sickle cell gene is prevalent within the Malaria belt and it is believed that sickle cell **trait** is protective against malaria.

Complications
Since SSD is a disease of the blood and the blood goes to all parts of the body, all parts of the body can be affected. In addition, while sickle cell trait has less complications than disease, it can, under certain circumstances, have all of the complications of sickle cell disease.

The complications from SSD arise when the red blood cells change shape (from a donut to a sickle shape) and become rigid. These cells then cause a cascade of events that then clog up (clot) the small and medium sized blood vessels subsequently starving the cells beyond the blockage of oxygen. When this happens, those cells die. Most of those cells do not regenerate and so **as a patient gets older more and more areas of the body die off until the organs fail. Therefore, SSD is a cumulative disease that get worse as you get older.**



The complications of SSD are too numerous to list here, but some of those that are more serious and potentially life threatening are listed.

Additionally, there has not been any steady decrease of hospitalization rates noted from 1998 to 2008 in sickle cell disease admissions, with only a slight decrease in length of hospital stay of 5.38 days in 1998 to 5.18 days in 2008
Renal Failure in Sickle Cell Disease: Prevalence, Predictors of Disease, Mortality and Effect on Length of Hospital Stay. Hemoglobin. 2016 Sep; 40(5): 295–299.

Pain (painful crises, VOC)

Pain results when cells are starved of oxygen and die. The larger the die off of cells the more severe the pain and the longer the duration. Bone pain is the most common. It is not that it feels like a broken bone, it is as if the bone is crushed and multiple fractures occur. It can cause severe and excrutiating pain.

The majority of medical contacts in sickle cell disease (SCD) are for exacerbations of pain due to vaso-occlusive episodes, commonly called “crises”.
Pain. 2009 September ; 145(1-2): 246–251.

Adult respondents in the Pain in Sickle Cell Epidemiology Study (PiSCES) reported SCD pain on 54.5% of the 31,017 days surveyed. Importantly, 29.3% of respondents had pain on greater than 95% of the days surveyed.

Those who described being in pain on 96% to 100% of days reported a mean pain intensity of 5.1 ± 0.2 on pain days and 6.2 ± 0.2 on crisis days, whereas those who described pain on 5% or fewer of days reported an intensity of 3.5 ± 0.4 on pain days and 4.5 ± 0.6 on crisis days. Opioid use was strongly correlated with pain intensity.

Pain in SCD is not only common, but also severe. Utilization due to SCD pain increased as patients grew older, from 0 to 30 years, and declined thereafter. Most SCD pain, even "crisis" pain, is managed at home, without emergency room or hospital utilization. The 8-state study reported a rate of **acute care encounters, 2.59 per patient per year. Re-hospitalization rates for the 8-state study were frequent: 22.1% and 33.4% at 14 and 30 d, respectively.**

Impacts of SCD Pain: on Depression, Psychological, Neurological Impacts, health related quality of life and sleep

ASH Education Book, **December 4, 2010** vol. 2010 no. 1 **409-415**

People with SCD may face long wait times before seeing a health provider in the ED and before receiving appropriate medicines.[1,2]Health providers in the ED may hold inaccurate beliefs about patients with SCD. Research shows that ED providers may suspect patients with SCD to be drug-seeking when they arrive in the ED.[3,4]

1. Haywood C Jr, Tanabe P, Naik R, Beach MC, Lanzkron S. The impact of race and disease on sickle cell patient wait times in the emergency department. Am J Emerg Med. 2013 Apr;31(4):651-6.
2. Tanabe P, Myers R, Zosel A, Brice J, Ansari AH, Evans J, Martinovich Z, Todd KH, Paice JA. Emergency department management of acute pain episodes in sickle cell disease. Acad Emerg Med. 2007 May;14(5):419-25.
3. Shapiro BS, Benjamin LJ, Payne R, Heidrich G. Sickle cell-related pain: perceptions of medical practitioners. J Pain Symptom Manage. 1997 Sep;14(3):168-74.
4. Waldrop RD, Mandry C. Health professional perceptions of opioid dependence among patients with pain. Am J Emerg Med. 1995 Sep;13(5):529-31.

SCD patients experienced wait times 25% longer than the General Patient Sample, though this difference was explained by the African-American race of the SCD patients. SCD patients waited 50% longer than did patients with long bone fracture even after accounting for race and assigned triage priority.

Haywood C Jr, Tanabe P, Naik R, Beach MC, Lanzkron S. The impact of race and disease on sickle cell patient wait times in the emergency department. Am J Emerg Med. 2013 Apr;31(4):651-6.

All of this is made more difficult by the fact that sickle cell patients, adult primarily, cannot find/get the specialized care that they need to prescribe pain medication and to prescribe adequate pain medication. The "opiod crisis" is preventing sickle cell patients, and all chronic pain patients, from getting appropriate care. Remember most sickle cell patients treat their pain at home. With inadequate pain treatment more and more of them will have to seek care from ERs.

If a chronically ill patient comes into an ER, such as an insulin dependent diabetic, and they know how much insulin they should be getting, they are considered a good patient. However, when a sickle cell disease patient with chronic pain comes into an ER and knows how much pain medication they need to control their pain, they are considered drug seekers/addicts and are usually ignored and not given appropriate medication for their pain. This results in inadequate pain control and then the patient

needs to be admitted to the hospital rather than discharged home with appropriate medication to get them over their pain crisis.

Stroke/Silent Stroke

Incident ischemic stroke was more frequent among those with SCT (13%) than those with homozygous hemoglobin A (10%).
Melissa C. Caughey, Laura R. Loehr, Nigel S. Key, Vimal K. Derebail, Rebecca F. Gottesman, Abhijit V. Kshirsagar, Megan L. Grove, and Gerardo Heiss. Stroke October 2014 Vol 45, Issue 10

Sickle cell trait may not be associated with incidence of ischemic stroke among African Americans. JAMA Neurol. 2018 Jul 1;75(7):802-807

The risk (for stroke) is enormous in SCD. Approximately 11% of SCD patients have clinically apparent strokes before the age of 20.**9** That risk increases to 24% by the age of 45.

The ischemic variant, which constitutes 54% of all cerebrovascular accidents (CVAs),[9]is **highest during the first decade and after age 30. During the 20s, ischemic CVA is replaced by hemorrhagic CVA.**[9] Although not characterized as age-dependent, 10% to 30% of SCD patients have silent strokes that exhibit radiologic findings consistent with diffuse white matter disease.[12–15] These silent infarcts are associated with cognitive deficiencies.[14]

Sickle cell disease and stroke, Luis A. Verduzco and David G. Nathan, Blood 2009 114:5117-5125

High prevalence of silent cerebral infarcts[4] and their association with lower IQ,[8,15] poor academic performance,[7] and increased risk for stroke.

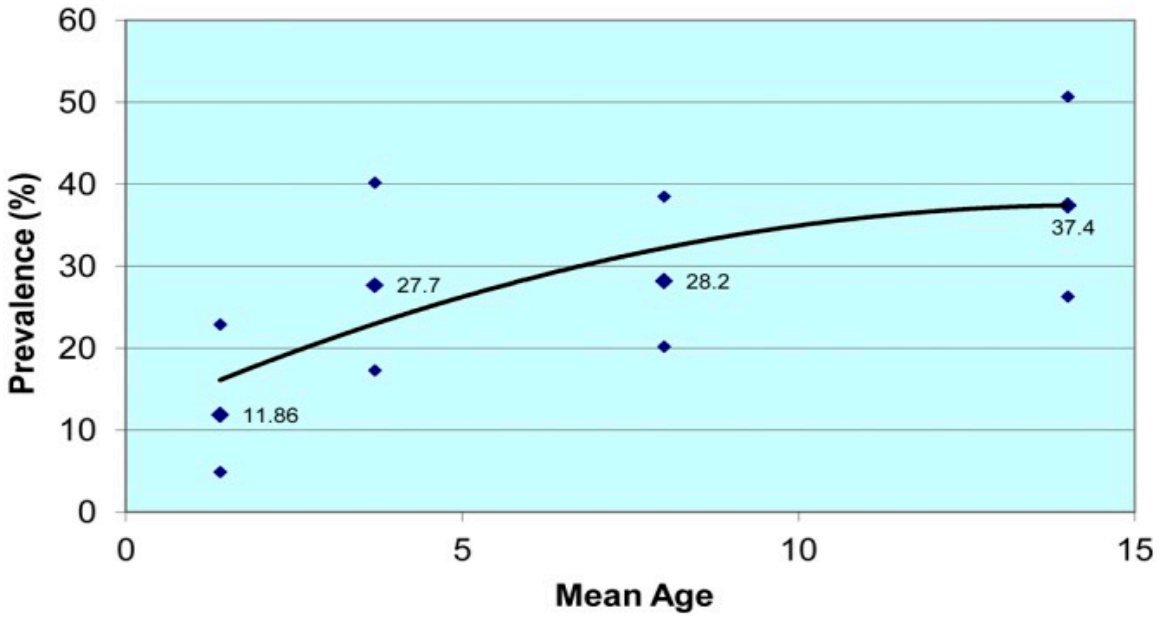
Silent Cerebral Infarct is defined as abnormal magnetic resonance imaging (MRI) of the brain in the setting of a normal neurologic examination without a history or physical findings associated with an overt stroke.

In a small study, the prevalence of silent cerebral infarcts at an average age of **13.7 months** was **13%.**[22] In a second study, in which surveillance MRI was conducted among children **up to 6 years** of age, the prevalence of silent cerebral infarct was **27%**.[23] In a third study, the prevalence **by 14 years** of age was **37%**.[4] Thus, the majority of silent cerebral infarcts have occurred in children with sickle cell anemia by 6 years of age.

AGE	% with Silent Stroke
13.7 months	13%
Up to 6 years	27%
By 14 years	37%
Up to 36 years	45%

Controlled Trial of Transfusions for Silent Cerebral Infarcts in Sickle Cell Anemia. N Engl J Med 2014; 371:699-710

Figure 5



Prevalence of SCI with 95% CIs plotted against age from 4 studies.[1,2,23,24]
Blood. 2012 May 17; 119(20): 4587–4596.

Among adults with SCA 43% had SCI at baseline. Of participants with baseline SCI, **30% had new or progressive SCI over 2.5 years** compared to 6% with no SCI at baseline

Silent infarct is a risk factor for infarct recurrence in adults with sickle cell anemia
Neurology August 21, 2018; 91 (8)

Silent cerebral infarctions are common in adults with SCD. Silent cerebral infarcts were present in 45% and overt strokes had occurred in 13% of adults with SCD.

Silent Cerebral Infarcts and Cerebral Aneurysms Are Prevalent in Adults with Sickle Cell Disease
Adetola A. Kassim, Sumit Pruthi, Matthew Day, Michael R. DeBaun and Lori C. Jordan
Blood 2014 124:2712;

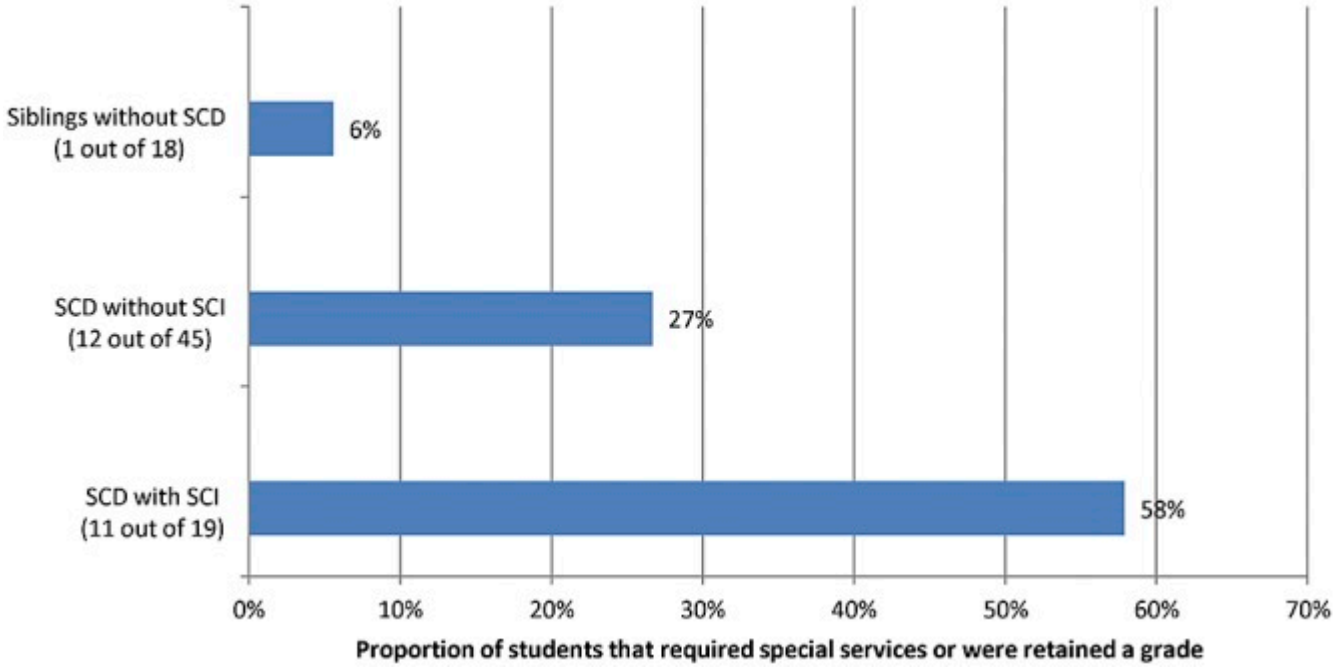
SCI is a risk factor for clinical stroke (14 fold higher) and progressive SCI

Children with SCI have lower cognitive test scores compared with children with a normal MRI of the brain. Poorer global intellectual function[17,22,26–28] has been reported in several studies, with function below the average range for the general population, but better than that of children with overt strokes. In a summary of global intelligence quotient in children with SCA and controls, Hogan et al graphically displayed data from multiple studies that included the Full Scale IQ (FSIQ) in controls without SCA, children with SCA with or without SCI, and children with SCA and overt stroke.[29] The gradient in FSIQ demonstrated the following consistent pattern: ethnically matched control children without SCA had a mean FSIQ greater than children with SCA and without SCI, who in turn had a FSIQ greater than those with SCI and covert and overt strokes

Specific areas of deficit have been associated with SCI, including executive functions like selective attention, card sorting, working memory, and processing speed,[4,30–32] visual motor speed and coordination,[4,22] vocabulary,[17,22,28,33] visual memory,[34] and abstract reasoning and verbal comprehension.[17,35] As a consequence of these specific deficits, academic achievement in math and reading are also affected, with one study reporting that the 35% of children with SCA and SCI had twice the chance of academic difficulties as those without SCI.[26]

SCI are associated with a specific cognitive profile correlating with their distribution in the frontal lobe, and are associated with cognitive deficits and academic difficulties

Figure 7



The proportion of students with SCA with and without SCIs and sibling controls that have either failed a grade or received special services.[26]

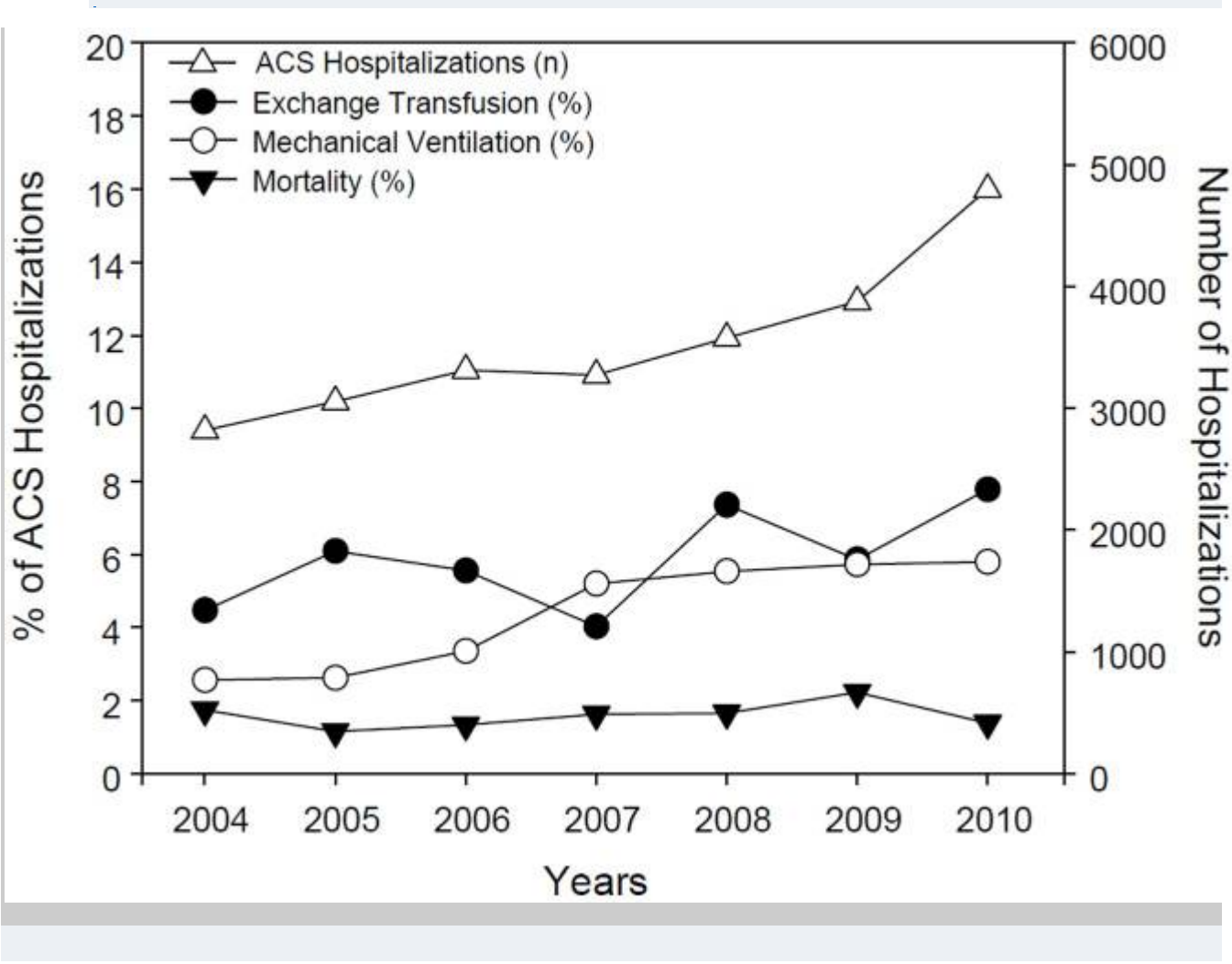
Silent cerebral infarcts: a review on a prevalent and progressive cause of neurologic injury in sickle cell anemia. Blood. 2012 May 17; 119(20): 4587–4596.

Acute Chest Syndrome

ACUTE CHEST SYNDROME (ACS) is a frequent complication of sickle cell disease (SCD) in patients hospitalized with vaso-occlusive crisis (VOC). It is associated with a high risk of sickle cell-related mortality and morbidity in children, including prolonged hospitalization. More than half of all children with homozygous SCD (HbSS) experience at least one episode of ACS in the first decade of life.[1] Recurrent episodes may herald the onset of debilitating chronic lung disease.[2]

The highest incidence of ACS is in children <10 years of age[36] with frequently occurring triggers in the form of pulmonary infections. Older children and adults more frequently present with dyspnea (labored breathing) and chest pain and tend to follow a more severe course.

Nearly half of all ACS episodes occur between 1 and 3 days after admission for severe VOC. Neurological complications, such as infarctive stroke, silent cerebral infarcts, and posterior reversible leukoencephalopathy syndrome, have been shown to be higher after severe episodes of ACS in children
Acute Chest Syndrome in Children with Sickle Cell Disease. Pediatr Allergy Immunol Pulmonol. 2017 Dec 1; 30(4): 191–201.



Acute Chest Syndrome Hospitalizations and Outcomes per year.

The descriptive figure shows the number of acute chest syndrome hospitalizations per year- identified by "n" (2004 to 2010)- Right y axis. Outcomes such as Exchange transfusion (%), Mechanical Ventilation (%) and Mortality(%) are shown as percentages of acute chest syndrome hospitalizations.(2004 to 2010)- Left y axis.

Close to 95.5% of all hospitalizations occurred on an emergent or urgent basis. About 29.6% of the hospitalizations were covered by Medicare, 40.5% by Medicaid, 20.2% by private insurance, and 3% by other insurance plans. About 6.7% were uninsured.

The mean hospital LOS was 7.8 days.
Outcomes of acute chest syndrome in adult patients with sickle cell disease: predictors of mortality. PLoS One. 2014 Apr 16;9(4):e94387

Kidney Failure
Renal involvement contributes substantially to the diminished life expectancy of patients with SCD, accounting for 16–18% of mortality. **Once ESRD (end stage renal disease) is reached, the mortality of patients who are on haemodialysis and have SCD is increased severalfold.**

As improved clinical care promotes survival into adulthood, SCN imposes a growing burden on both individual health and health system costs.

Proteinuria (protein in the urine) occurs in up to 27% of patients in the first three decades,[15,58] and in up to 68% of older patients.[15,59]

In a study that involved 98 patients over 5 years the prevalence of CKD (chronic kidney disease) rose from 29% to 42% over this period.

Prior studies demonstrate that irreversible kidney damage (defined by a serum creatinine level >132.6 μmol/l) occurs in approximately 12% of patients with SCD.[62]

In SCT (sickle cell trait), only 40% of RBC haemoglobin content is HbS: the rest is normal HbA. Nevertheless, this amount of HbS is sufficient to cause common complications such as haematuria (blood in the urine) and impaired concentrating ability. A 2014 analysis demonstrated that SCT is clearly associated with an increased risk of CKD and a reduction in GFR.
Sickle cell disease: renal manifestations and mechanisms. Nat Rev Nephrol. 2015 Mar; 11(3): 161–171.

Both ARF (acute renal failure) and CKD (chronic kidney disease) were associated with higher risk of inpatient mortality, longer duration of the hospital stay and expensive hospitalizations. The yearly incidence of new ARF in sickle cell disease patients was 1.4% and annual CKD incidence was 1.3%.

the prevalence of CKD in adults with sickle cell disease was 5.0% and ARF was 4.0%, and incidence of both almost tripled compared to adults without sickle cell disease. Both CKD and ARF conditions were associated with higher mortality and health care utilization in this study.

in a prospective trial that enrolled 725 patients with sickle cell disease, 4.2% developed renal failure, with a median survival of 4 years and **median age at diagnosis of 23 years. The incidence of renal failure increased to 12.0%, with a median age at diagnosis of 37 years.**

Both ARF and CKD were associated with increased mortality during hospital admission. There was a 3.6 relative risk (RR) for death on admission with CKD (95% CI: 2.6–5.0) and an even higher 9.5 RR for death on admission with ARF.

It is now evident that age is a predictor of developing renal failure in sickle cell disease.
Renal Failure in Sickle Cell Disease: Prevalence, Predictors of Disease, Mortality and Effect on Length of Hospital Stay. Hemoglobin. 2016 Sep; 40(5): 295–299.

Life Expectancy

For HIV
This means that a 20 year-old person living with HIV in these regions (including the US), starting treatment after 2008, can now expect to live to 78 (vs life expectancy of 54 years for sickle cell disease).

https://www.avert.org/news/life-expectancy-people-hiv-now-near-normal-%E2%80%93-only-those-accessing-treatment accessed 1.25.19

For Sickle Cell Disease

The median age of death for sickle cell disease in the US has decreased from 1994 – 2005. A decrease in median death of 4 years for males and 6 years for females. Life expectancy can be improved/increased with optimal management of their disease.

For SS patients the median age of death for males was 42, for females was 48 in 1994

Mortality in sickle cell disease. Life expectancy and risk factors for early death.

Platt OS[1], Brambilla DJ, Rosse WF, Milner PF, Castro O, Steinberg MH, Klug PP.

N Engl J Med. 1994 Jun 9;330(23):1639-44.

Median age of death for males in 2005 was for males 38, for females 42

Mortality Rates and Age at Death from Sickle Cell Disease: U.S., 1979–2005
Sophie Lanzkron, MD, MHS, C. Patrick Carroll, MD, Carlton Haywood, Jr., PhD, MA
Public Health Rep 2013;128(2):110–116

In NYS, from 2004-2008, only 14% of sickle cell disease patients were 51 years or older.
https://www.cdc.gov/ncbddd/sicklecell/documents/SCD_in_NY_Prov.pdf

Cost of Disease

For an average person with SCD reaching age 45, total lifetime health care costs were estimated to be nearly $1 million, with annual costs ranging from over $10,000 for children to over $30,000 for adults.

Teresa L. Kauf, Thomas D. Coates, Liu Huazhi, Nikita Mody-Patel and Abraham G. Hartzema,
"The cost of health care for children and adults with sickle cell disease,"
American Journal of Hematology 84, no. 6 (March 2009): 323-327.

The most recent data available shows that costs for hospital stays due to sickle cell disease complications were estimated at $488 million (2004)
https://www.cms.gov/About-CMS/Agency-Information/OMH/about-cms-omh/blog/sickle-cell-disease-care.html accessed 1.25.19

SCD is a major public health concern. From **1989 through 1993**, an average of 75,000 hospitalizations due to SCD occurred in the United States, costing approximately $475 million.
https://www.cdc.gov/ncbddd/sicklecell/data.html accessed 1/25/19
Obviously, costs have increased since then and, as noted below, are now about $1Billion
Sickle Cell Disease is the most costly disease/patient to NYS Medicaid

Sickle cell disease costs~$15,000/patient while the next costly disease/patient is HIV at ~$10,000/patient. Therefore, sickle cell disease costs ~ 50% more/patient than HIV
Personal communication

Other states will a smaller population of sickle cell disease spend more on sickle cell disease care than NYS.

North Carolina	**>$4M**
Pennsylvania	**$1.26M**
Illinois	**$500,000 for one sickle cell program**

NYS has cut funding for care of sickle cell disease patients over the last 20 years by about 66% ($500,000 to $170,000)

With a decrease in cost of ~3.3%/patient to NYS Medicaid, NYS Medicaid could save between ~$4-$5M/year.

For hospital stays primarily due to SCD, "66% were paid by Medicaid and 13% were paid by Medicare
The cost of hospitalizations for treating acute pain alone is now estimated at ~$1 billion

https://www.forbes.com/sites/judystone/2015/06/19/sickle-cell-disease-highlights-racial-disparities-in-healthcare/#3da803723b75 access 1/25/19

Sickle Cell patients on Medicaid receive poorer care than those on commercial insurance.

Use of hematology/oncology care was strikingly low among Medicaid SCD patients. This finding suggests that SCD patients in Medicaid plans may have less access to hematologists/oncologists than patients with commercial insurance. This limited use of specialty care may reduce the preventative care Medicaid patients receive. The higher ED and inpatient use and lower HU compliance in the Medicaid population may be indicative of greater severity and/or unmet need.

Access to Care for Medicaid and Commercially-Insured United States Patients with Sickle Cell Disease

Carlton Dampier, Julie Kanter, Robin Howard, Irene Agodoa, Sally Wade, Virginia Noxon and Samir K. Ballas Blood 2017 130:4660;

Costs to community hospitals is a burden
40.5% having Medicaid, and 54.1% with Medicare

As healthcare costs continue to be scrutinized, a more conscious effort will need to be placed on delivering high quality cost-effective care to our sickle cell population. From this analysis, there is a clear economic burden of sickle cell related hospitalizations to community hospitals. It is also clear that there is a small subset of patients who consume a large percentage of the resources. This may lend itself well to focused collaborative care management services of these high consumers of healthcare resources.

The inpatient management of sickle cell vaso-occlusive crisis is well known, but the goal of treatment extends beyond that of just inpatient management. Patients with SCD need effective management in the outpatient setting in hopes to prevent readmissions, reduce hospital length of stays, and ultimately decrease the economic burden to our healthcare system.

Economic Impact of Sickle Cell Hospitalization: Rahul Singh, Ryan Jordan and Charin Hanlon Blood 2014 124:5971

Lack of funding for sickle cell disease

Average annual NIH funding per affected individual was 3.4-fold greater for CF than SCD from 2008 to 2016. Between 2008-2012, private foundation funding was 161-fold greater for CF than SCD. Between 2013-2016, private funding was 971-fold greater for CF than SCD. There were 1.8 times as many PubMed publications for CF compared to SSD.

Table I: Funding and Research Output for Sickle Cell Disease (SCD) and Cystic Fibrosis (CF)

DISEASE CHARACTERISTICS	Sickle Cell Disease (SCD)		Cystic Fibrosis (CF)	
Prevalence (USA)	90,000		30,000	
Estimated new cases annually (Global)	300,000		1,000	
Average Life Span (years)	48		41	
AVERAGE ANNUAL FUNDING	**2008-2012**		**2013-2016**	
	SCD	CF	SCD	CF
NIH Funding	$69 million	$85 million	$78 million	$81 million
NIH Funding per individual affected	$769	$2847	$867	$2700
Foundation Revenue	$6.4 million	$342 million	$6.4 million	$2.2 billion
Foundation Revenue per individual affected	$71	$11,420	$72	$69,177
RESEARCH OUTPUT	**2008-2012**		**2013-2017**	
	SCD	CF	SCD	CF
Average Annual PubMed Publications	799	1505	1049	1856
Total Interventional Clinical trials	92	128	137	130
Average Annual Total Trials	18.4	25.6	27.4	26
Average Annual NIH-Federal Funded	5.4	2.2	5	1.2
Average Annual Industry Funded trials	6.8	14.4	6.4	15.4
Average Annual university/philanthropic funded trials	6.8	9.6	16.6	9.4
New FDA Drug Approvals	0	2	1	2
New FDA Drug Indications	0	6	2	5

Disparities in Foundation and Federal Support and Development of New Therapeutics for Sickle Cell Disease and Cystic Fibrosis. Faheem Farooq, MD, MPH[1*] and John J Strouse, MD, PhD. ASH Poster 2018

Little Data on Sickle Cell Disease and Sickle Cell Trait in NYS

There is shockingly little data on sickle cell epidemiology in the U.S., since there is very limited national surveillance data, explained Suzette Oyeku, MD, MPH, a sickle cell expert and health services researcher at The Children's Hospital at Montefiore/Albert Einstein College of Medicine. She also stressed that the transition time from pediatrics to adult care "is a critical time period. The risk of early death increases in this time frame."
https://www.forbes.com/sites/judystone/2015/06/19/sickle-cell-disease-highlights-racial-disparities-in-healthcare/#3da803723b75 accessed 1/25/19

1. How many sickle cell disease patients are currently residing in NYS
2 How many trait patients currently reside in NYS?
3 How many sickle cell disease patients are not in care – ie can't find a primary physician or primary hematologist to take care of them
4 How well do community hospitals do with the care of sickle cell disease patients?
5 How much does it cost to take care of a sickle cell disease patient?
6 How much are hospitals and physicians actually reimbursed to take care of sickle cell disease patients?

7 How can we educate communities, particularly immigrant communities, about sickle cell trait/disease?
8 How can we educate healthcare professionals to treat sickle cell disease patients with respect and dignity?
9 How do we erase the stigma of sickle cell disease?
10 How do we erase the healthcare disparities for sickle cell disease patients?
11 How do we get hospital administrators to prioritize the health and well being of sickle cell patients?

The department of health has many programs, but to avail yourself of a program and you have to be in the care of a healthcare provider. Since most sickle cell disease patients avoid healthcare institutions because of the bias they receive most patients/programs don't know of available programs.

Montefiore had a program for a day hospital for sickle cell disease patients in the 90s and early 2000s. It was subsidized by being one of 10 national comprehensive sickle cell centers. Dr. Benjamin reviewed data before the hospital was opened and compared to 5 years after. Patients, usually with painful crisis could go to the day hospital, when beds available, instead of the ER. In the year prior to the day hospital opening 92% of patients were admitted to the hospital, while in the last 3 years only 6-10% were admitted from the day hospital. With the reduction in admissions and decrease in LOS, it was estimated to save $1.7 million (much more in today's dollars) in addition to providing superior care/outcomes.
Sickle cell anemia day hospital: an approach for the management of uncomplicated painful crises. Blood 2000 95:1130-1136;

Yet this day hospital was closed by hospital administration shortly after the Federal Government stopped funding the national comprehensive sickle cell centers despite the outcry from patients. Why would this program close when it clearly provided better care to patients in painful crisis and saved money?

Sickle cell disease patients require much support to avail themselves of the healthcare system. Most patients are lower socioeconomically and so have difficulty with transportation to many clinic visits, are cognitively impaired by the silent strokes and so have difficulty with memory and problem solving to get to appointments, and frequently do not have adequate availability of social work staff to help them navigate these myriads of problems.

Dr. Kato from Pittsburgh has an anecdote about an adult patient who was scheduled to see a kidney specialist. The patient arrived in time for his appointment but was so confused about the maze of buildings and could not problem solve how to get to his appointment. He gave up, went home and missed his appointment. The cognitive deficits affecting the health of sickle cell patients are very real and very underappreciated.

Mental health issues that arise from having a debilitating chronic illness coupled with brain dysfunction and deficits is also a big issue for sickle cell disease patients and usually not very well addressed.

The previous sickle cell bill submitted to the legislature would address many of the issues above, and it is hoped the new bill will as well. Funding is needed to improve the care of patients with sickle cell disease/trait NOW and to formally collect data, so desperately needed, to better understand and address the healthcare disparities that sickle cell disease patients endure in NYS.