Sickle Cell Disease Fact Sheet

What is SCD?

- Sickle cell disease (SCD) is a rare, genetic, life-shortening blood disorder that affects every organ in the body.
- Normal red blood cells are round and flexible, which lets them travel through small blood vessels to deliver oxygen to all parts of the body. In people with with sickle cell, red blood cells form into a crescent, sickle shape that break apart easily, clump together, and block the flow of blood. This can cause a range of serious health issues.

SCD in the US

- Roughly 100,000 people have sickle cell disease in the U.S, although comprehensive surveillance and reporting is lacking and the exact number of cases in the US is unknown. Citation
- 1 out of 365 Black or African American babies in the U.S. are born with SCD. Citation
- There are almost 42,000 Medicaid and CHIP beneficiaries with SCD. In 2017, 49% of them had at least one hospital stay. Citation

Symptoms and Complications

- SCD is a complex disease and the types and severity of symptoms can differ widely from person to person.
- Recurrent pain crisis is one of the most common symptoms of SCD. Some people say the pain associated with SCD is on the same pain level as gunshot and stab wounds. When an adult with SCD has a pain crisis, it can last 9 to 11 days on average. Citation
- Chronic complications can occur across multiple organs and include death of bone tissue, skin ulcers, stroke, lung damage, blindness, neurocognitive impairment, pulmonary hypertension, heart and kidney failure, and can result in early mortality.

Treatment

- Because the type and severity of SCD symptoms can differ widely from person to person, treatment plans should be tailored to individual's unique circumstances.
- Until recently only three specific interventions were considered helpful for SCD: stem cell transplantation, chronic transfusions, and hydroxyurea.
- The FDA recently approved three transformative new treatments for sickle cell disease, the first in 20 years.
- While sickle cell disease has been neglected for far too long, today there are many, many reasons for hope as new curative therapies are in the pipeline.

All-Encompassing Condition

- The impact of SCD on quality of life is complex and affects family and caregivers in many ways.
- The lack of treatment options, discrimination, stigma around the need for chronic pain management, disruption of family and social activities, missed school and/or work all combine to make living with SCD very difficult. Citation
- SCD presents challenges at home, school, work, and social relationships.
- People with SCD often end up on formal disability programs, which unfortunately carries its own stigma.

Access to Care

- Only one in four patients with sickle cell disease receives standard of care.
- All SCD patients experience challenges with access, quality, and affordability of care.
- People with SCD have less access to comprehensive team care than people with genetic disorders such as hemophilia and cystic fibrosis. Citation
- The picture of "baseline" or "usual" care for patients with SCD is highly variable. Deep dysfunction in care is driven by poor coordination within provider systems and by many others barriers to access.

Cost of SCD

- This disease is costly; expenditures for patients with SCD are six times higher than non-SCD patients on Medicaid and 11 times higher than non-SCD patients with private insurance. Citation
- The total health system economic burden of SCD estimated at $2.98 billion per year in the US with 57% due to inpatient costs, 38% due to outpatient costs, and 5% due to out-of-pocket costs. Citation
- Sickle cell disease management can become very expensive, costing families thousands a year in out-of-pocket costs.

Economic Impact

- Significant losses in work productivity for patients and caregivers contribute to the burden.
- Patients with more frequent or severe pain crises are more likely to have experienced losing a job or having to reduce work hours due to their SCD. Citation

Racism and Stigma

- African Americans are disproportionately affected by SCD.
- Stigma of SCD is a pressing health concern. Factors that contribute to stigma in SCD include racism, pain and opioid use, disease severity, and sociodemographic characteristics. Citation
- Patients with SCD presenting to an emergency room for care experience longer wait times than other groups, and are often falsely accused of exaggerating their pain or seeking drugs. Citation

Public Interest

- In 2018, the President signed into law a bipartisan bill that reauthorizes a sickle cell disease prevention and treatment program and grants for research, surveillance, prevention, and treatment of heritable blood disorders. Citation
- HHS has set the goal of extending the lives of Americans with sickle cell disease by 10 years within 10 years through action like the Health+ initiative. Citation
- Our SCD community has come a long way in igniting public interest to make sickle cell a public health concern, but we're not done! We are working every day to build on this incredible momentum, and we won't stop until there is a cure for every person with SCD.