

[Calling for Sickle-Cell Trait Research, Surveillance, and Public Education and Awareness \(H Res 539, 115th Congress\)](#)

Encourages increased awareness and research support for sickle-cell disease and promotes standardized strategies for sharing education, screening results, and counseling.

Updated last **May 31, 2018**

for the 09/26/2017 version of H Res 539.

WHAT IT DOES

Calling for Sickle-Cell Trait Research, Surveillance, and Public Education and Awareness ([H Res 539](#)) aims to improve the overall awareness of sickle-cell disease (SCD). The nonbinding resolution recommends implementation of standardized protocols both for raising awareness of SCD and for dissemination of information, including screening results and counseling, to those carrying or affected by the disease.

This resolution specifically:

- Recognizes the challenges of treating health outcomes associated with SCD;
- Encourages the medical community to work with states' [newborn screening programs](#) to standardize the dissemination of screening results, education, and counseling regarding SCD;
- Urges the Department of Human and Health Services (HHS) to develop a public awareness campaign that promotes the importance of knowing one's sickle-cell trait status no matter one's racial or ethnic identity;
- Calls on HHS to expand access for screening and counseling for individuals with the sickle-cell trait; and
- Supports research concerning outcomes and implications SCD.

RELEVANT SCIENCE

According to the National Institutes of Health's [National Heart, Lung, and Blood Institute](#), sickle-cell disease (SCD) encompasses a group of genetic disorders affecting red blood cells (RBCs) in which the individual has abnormal or sickle-shaped RBCs that cannot efficiently transport oxygen throughout the body.

All tissues in the body need a steady supply of oxygen to function correctly. In order to reach all parts of the body, oxygen-carrying RBCs need to squeeze in and out of tight blood vessels. [Hemoglobin](#), the protein in RBCs that carries and delivers oxygen, typically unloads its oxygen in various body tissues and then travels back into the RBCs to get reloaded with oxygen in the lungs.

Red blood cells normally have a flexible donut shape which allows them to travel through small blood vessels. But sickled hemoglobin form stiff rods inside these cells resulting in their deformation. Thus, passage through the circulatory system is restricted, resulting in reduction of oxygen delivery throughout the body.

[The Mayo Clinic](#) outlines the following common symptoms of SCD: painful swelling of hands and feet, frequent infections due to damage to the spleen (the organ that fights infection), yellow tint to the skin and eyes (jaundice), fever, and abdominal swelling. Later in life, those affected by SCD are at risk for stroke, hypertension, blindness, ulcers, and gallstones.

Sickle cells also tend to burst and die more frequently than normal cells, leading to a reduction in RBC count, indicative of a condition called [hemolytic anemia](#). Symptoms of hemolytic anemia include severe fatigue, stress on many organs (such as the lungs, heart and kidneys,) and other severe complications involving the lungs, spleen, eyes, joints, and skin.

Sickle-cell disease is not contagious, meaning it does not spread laterally from person to person as does the common cold. Rather, SCD is inherited, meaning the genes causing it are passed from parent to offspring. Sickle-cell genes are [autosomal recessive](#), meaning that you must inherit a sickle-cell allele from each parent in order to develop the most severe form of SCD, sickle cell anemia. However, inheritance of one sickle cell [allele](#) (from only one parent) results in carrier status, meaning there is a 50% chance of the SCD trait being passed to the next generation.

Additionally, one copy of the SCD allele is sufficient to cause the body to produce some abnormal hemoglobin along with normal hemoglobin. This is referred to as [sickle-cell trait](#) (SCT). People with SCT usually do not present symptoms of a disease, but may be at increased risk for complications in a variety of extreme environmental conditions such as high atmospheric pressure, low oxygen levels, and high altitude. Athletes with SCT are especially at risk when undergoing training and conditioning exercises under severe conditions due to an increased risk for heat stroke and muscle breakdown.

Sickle-cell disease and sickle-cell trait can both be diagnosed by a blood test. According to the [Centers for Disease Control and Prevention](#) and the [Sickle Cell Disease Association of America](#), approximately 70,000 – 100,000 people are affected by SCD in the United States, most commonly African American and Hispanic American individuals. Approximately 7% of African American babies have SCT.

Sickle-cell disease affects people throughout their lives; the only current cure is [hematopoietic stem cell](#) transplantation, a costly treatment that depends on the availability of donors who are a close enough genetic match to a given patient. However, if the disease is caught early enough, there are a number of treatments available that will extend life expectancy and improve quality of life. The implementation of these treatments [has increased life expectancy](#) from fourteen years in 1973 to approximately 50 years today.

Recent research has looked at the possibility of a vaccine that protects against pneumococcal disease to decrease mortality rates among children. [One study](#), published in 2009, found that the introduction of the vaccine led to a 42% decrease in mortalities among African American babies under four years old born with SCD. [Another study](#) found similar results in regards to children under four, but did not find a significant decrease in mortality rates for children between the ages of four and fourteen. The researchers of this latter study concluded that more research needs to be done in order to develop more effective treatment approaches, such as [hydroxyurea](#), for older children to significantly decrease mortality rates in this age demographic.

RELEVANT EXPERTS

[Marilyn Jo Telen, MD](#) is a Professor of Medicine, an Associate Professor of Pathology, and a member of the Duke Cancer Institute at Duke University. Her research and practice to date has been focused on the molecular genetics and biochemistry of blood antigens and SCD. Dr. Telen is also the Director of Duke's Comprehensive Sickle Cell Center. Her work and publications focus on the composition of red blood cell membranes and she is also involved in a large study looking for genetic predisposition to development of SCD.

Relevant publications:

- Kanter, Julie, Marilyn J. Telen, Carolyn Hoppe, Christopher L. Roberts, Jason S. Kim, and Xiaoxi Yang. 2015. "Validation of a Novel Point of Care Testing Device for Sickle Cell Disease." *BMC Medicine* 13: 225. <https://doi.org/10.1186/s12916-015-0473-6>.
- Elmariah, Hany, Melanie E. Garrett, Laura M. De Castro, Jude C. Jonassaint, Kenneth I. Ataga, James R. Eckman, Allison E. Ashley-Koch, and Marilyn J. Telen. 2014. "Factors Associated with Survival in a Contemporary Adult Sickle Cell Disease Cohort." *American Journal of Hematology* 89 (5): 530-535. <https://doi.org/10.1002/ajh.23683>.

[John J. Strouse, MD](#) is an instructor in the Department of Medicine at Duke University. Dr. Strouse's work is centered around pediatric hematology and developing new strategies and therapies for treating people with SCD.

Relevant publications:

- Stewart, Rosalyn W., Lauren N. Whiteman, John J. Strouse, C. Patrick Carroll, and Sophie Lanzkron. 2016. "Improving Inpatient Care for Individuals with Sickle Cell Disease Using the Project ECHO Model." *Southern Medical Journal* 109(9): 568-569. <https://doi.org/10.14423/SMJ.0000000000000509>.
- Noronha, Suzie A., S. Christy Sadreameli, and John J. Strouse. 2016. "Management of Sickle Cell Disease in Children." *Southern Medical Journal* 109(9): 495-502. <https://doi.org/10.14423/SMJ.0000000000000523>.
- Strouse, John J. 2016. "Hydroxyurea for Sickle Cell Disease: Now Is the Time!" *Southern Medical Journal* 109(9): 535-537. <https://doi.org/10.14423/SMJ.0000000000000515>.

[Nirmish Ramesh Shah, MD](#) is an Assistant Professor in the Department of Medicine and Pediatrics at Duke University. Dr. Shah's work focuses on understanding the biology of SCD as well as pain management, symptom analysis, and hospitalization strategies for those affected by this disease.

The diagnosis of sickle trait is greatly under-appreciated for its implications towards future generations. Due to its implications, a concerted effort is needed to assure the diagnosis of sickle cell trait follows patients as they grow up and move from pediatrics to adult medical care.

Relevant publications:

- Lunyera, Joseph, Charles Jonassaint, Jude Jonassaint, and Nirmish Shah. 2016. "Attitudes of Primary Care Physicians Toward Sickle Cell Disease Care, Guidelines, and Comanaging Hydroxyurea with a Specialist." *Journal of Primary Care & Community Health* 8(1): 37-40. <https://doi.org/10.1177/2150131916662969>.
- Shah, Nirmish, Jude Jonassaint, and Laura De Castro. 2014. "Patients welcome the Sickle Cell Disease Mobile Application to Record Symptoms via Technology (SMART)." *Hemoglobin* 38(2): 99-103. <https://doi.org/10.3109/03630269.2014.880716>.
- Shah, Nirmish, Margon Rollins, Daneil Landi, Radhika Shah, Jonathan Bae, and Laura M. De Castro. 2014. "Differences in Pain Management Between Hematologists and Hospitalists Caring for Patients with Sickle Cell Disease Hospitalized for Vaso-Occlusive Crisis." *Clinical Journal of Pain*, 30(3): 266-268. <https://doi.org/10.1097/AJP.0b013e318295ec04>.

[Jennifer Rothman, MD](#) is an associate professor of Pediatrics and an Assistant Professor in Medicine at Duke University. Dr. Rothman's research and work centers around the development of SCD, bone marrow failure syndromes, and general hematology.

Relevant publications:

- Estep, Jeremie H., Chiara Melloni, Courtney D. Thornburg, Paweł Wiczling, Zora Rogers, Jennifer A. Rothman, Nancy S. Green, et al. 2015. "Best Pharmaceuticals for Children Act- Pediatric Trials Network Administrative Core Committee. Pharmacokinetics and Bioequivalence of a Liquid Formulation of Hydroxyurea in Children with Sickle Cell Anemia". *The Journal of Clinical Pharmacology* 56(3): 298-306. <https://doi.org/10.1002/jcph.598>.
- Luchtman-Jones Lori, Sara Pressel, Lee Hilliard, R. Clark Brown, Mary G. Smith, Alexis A. Thompson, Margaret T. Lee, et al. 2015. "Effects of hydroxyurea treatment for patients with hemoglobin SC disease". *American Journal of Hematology* 91(2): 238-242. <https://doi.org/10.1002/ajh.24255>.
- Ware Russell E., Barry R. Davis, William H. Schultz, R. Clark Brown, Banu Aygun, Sharada Sarnaik, Isaac Odame, et al. 2015. "Hydroxycarbamide Versus Chronic Transfusion for Maintenance of Transcranial Doppler Flow Velocities in Children with Sickle Cell Anaemia-TCD with Transfusions Changing to Hydroxyurea (TWITCH): A Multicentre, Open-Label, Phase 3, Non-Inferiority Trial". *The Lancet* 38, no. 10019: 661-670. [https://doi.org/10.1016/S0140-6736\(15\)01041-7](https://doi.org/10.1016/S0140-6736(15)01041-7).
- Wang, Hsin-Hsiao Scott, Katherine W. Herbst, Jennifer A. Rothman, Nirmish R. Shah, John S. Wiener, and Jonathan C. Routh. 2015. "Trends in Sickle-Cell-Disease-Related Priapism in U.S. Children's Hospitals". *Urology* 89: 118-122. <https://doi.org/10.1016/j.urology.2015.11.023>.
- Allen, Taryn M., Lindsay M. Anderson, Jennifer A. Rothman, and Melanie J. Bonner. 2016. "Executive Functioning and Health-Related Quality of Life in Pediatric Sickle Cell Disease". *Child Neuropsychology* 8: 889-906. <https://doi.org/10.1080/09297049.2016.1205011>.
- Cancio, Maria I., Banu Aygun, David H. K. Chui, Jennifer A. Rothman, J. Paul Scott, Jeremie H. Estep, and Jane S. Hankins. 2017.

"The Clinical Severity of Hemoglobin S/Black (Aγδβ)0-thalassemia". *Pediatric Blood & Cancer* 64(11).
<https://doi.org/10.1002/pbc.26596>.

BACKGROUND

All newborns in the US are currently required to be tested for SCD as part of the [newborn screening program](#) within each state, district, or territory. This test is done through a heel prick that collects a small blood sample on a special paper that can then be sent to a lab to detect the type of hemoglobin present in the sample. If a baby is positive for SCD, health providers [are instructed](#) to follow up with the family to make sure the parents are informed of the results. The child is then retested to confirm the diagnosis. These tests also inform the parents as to whether or not the child carries the SCT trait. Health providers are further encouraged to recommend counseling to these families and children.

It is also possible to [diagnose SCD and SCT prenatally](#) (i.e., before birth.) This diagnosis is done by taking a sample from the amniotic fluid (the liquid in the sac carrying the growing embryo) as early as eight to ten weeks into pregnancy. This diagnosis could also be done by taking tissue from the placenta, the organ that connects the umbilical cord to the womb.

ENDORSEMENTS & OPPOSITION

At present, there have not been any publicly reported endorsements of or opposition to this resolution.

STATUS

H Res 539 was introduced in the House on September 26, 2017 and referred to the [House Committee on Energy and Commerce](#). On September 29, 2017, it was referred to the [Subcommittee on Health](#).

RELATED POLICIES

- Sickle Cell Disease Awareness Month ([S Res 273](#)): Encourages further education in communities across the United States about SCD; and promotes the need for research, early detection methods, effective treatments, and preventative care programs with respect to SCD, complications from SCD, and conditions related to SCD. This resolution was agreed to in the Senate on September 28, 2017 by unanimous consent.
- Sickle Cell Disease Research, Surveillance, Prevention, and Treatment Act of 2017 ([HR 2410](#)) ([SciPol brief available](#)): This bill would authorize the Department of Health and Human Services to incentivize states through grants and funding to collect data on the prevalence and treatment of SCD and SCT as well as to conduct public health initiatives to improve the quality of care and treatment for those suffering from this disease. This bill passed the House by voice vote on February 26, 2018 and was referred to the Senate on the next day.

SPONSORS

Sponsor: [Representative Barbara Lee \(D-CA-13\)](#)

Cosponsors:

- [Representative Michael C. Burgess](#) (R-TX-26)
- [Representative Eleanor Holmes Norton](#) (D-DC-at large)
- [Representative Frederica S. Wilson](#) (D-FL-24)
- [Representative Bonnie Watson Coleman](#) (D-NJ-12)

- [Representative Raul M. Grijalva](#) (D-AZ-3)
- [Representative Dwight Evans](#) (D-PA-2)
- [Representative Alcee L. Hastings](#) (D-FL-20)
- [Representative Shelia Jackson Lee](#) (D-TX-18)
- [Representative Keith Ellison](#) (D-MN-5)
- [Representative Gregory W. Meeks](#) (D-NY-5)
- [Representative Donald M. Payne, Jr.](#) (D-NJ-10)

PRIMARY AUTHOR

Hannah Brown, PhD Candidate

EDITOR(S)

Allison Roder, PhD Candidate; Andrew Pericak, MEM

RECOMMENDED CITATION

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