2020 Sickle Cell Task Force Annual Report

As Required by

Texas Health and Safety Code,

Section 50.007

Sickle Cell Task Force

December 2020

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Executive Summary

Texas Health and Safety Code, Section 50.007 requires the Sickle Cell Task Force (Task Force) to submit an annual report to the Governor and the Legislature by December 1 of each year (starting December 1, 2020). The report is required to summarize the Task Force's work and include any recommended actions or policy changes endorsed by the Task Force.

The purpose of the Task Force is to study and advise the Department of State Health Services on implementing the recommendations made in the 2018 Sickle Cell Advisory Committee Report published by the Sickle Cell Advisory Committee (abolished on September 1, 2018) or any other report the Executive Commissioner determines is appropriate.

Major activities of the Task Force during fiscal year 2020 include the following:

- Appointing a presiding officer;
- Approval of Task Force bylaws;
- Meeting with subject-matter experts related to recommendations in the 2018 report;
- Forming subcommittees to address the 2018 recommendations at a higher level; and
- Developing milestones for each of the recommendations in the 2018 report.

Proposed activities of the Task Force for fiscal year 2021 include the following:

- Continuing to engage with subject-matter experts on Medicaid, sickle cell disease (SCD)/sickle cell trait (SCT) surveillance, and Community Health Workers;
- Creating SCD/SCT public awareness campaigns;
- Reviewing current SCD/SCT surveillance in the state and recommending possible enhancements to the current surveillance system;
- Reviewing current Continuing Education Units and State Module courses that provide SCD/SCT education;
- Reviewing current Medicaid contracts for improvements in medical treatments and services for SCD population; and

- Exploring ways to improve access points to health care, such as access to Sickle Cell medical homes.

1. Introduction

The Sickle Cell Task Force (Task Force) was established in accordance with House Bill 3405, 86th Legislature, Regular Session, 2019, and promulgated under the Texas Health and Safety Code, Chapter 50. Statute directs the executive commissioner of the Health and Human Services Commission to establish and maintain a task force to raise awareness of sickle cell disease and sickle cell trait. In August 2019, the Health and Human Services executive commissioner delegated the creation and maintenance of the Task Force to the Department of State Health Services (DSHS).

The purpose of the Task Force is to study and advise DSHS on implementing the recommendations made in the 2018 Sickle Cell Advisory Committee Report published by the Sickle Cell Advisory Committee or any other report the executive commissioner determines is appropriate.

The Task Force is required to prepare and submit to the governor and the legislature an annual written report that summarizes the Task Force's work and includes any recommended actions or policy changes endorsed by the Task Force no later than December 1 each year, beginning December 1, 2020.

In accordance with Texas Health and Safety Code, Chapter 50, this report outlines the following:

- A summary of the Task Force's actions during fiscal year 2020;
- A description of the Task Force's attendance (Appendix A), activities and costs during fiscal year 2020; and
- The Task Force's plans for future work.

2. Background

Sickle cell disease (SCD) was first described in the United States (US) in 1910 when "sickled" red blood cells were identified in an individual with anemia. Over the next century, the cause of the unusual shaped red blood cells was discovered as a single change in one of the genes that produces hemoglobin. By the 1970's, SCD was recognized as an inherited blood disease that primarily affected Black Americans and was associated with significant morbidity, mortality, and health care costs. In 1972, the National Sickle Cell Control Anemia Act was initially passed by Congress,[1] which led to subsequent legislation efforts as recent as 2018. See Appendix B for a timeline of legislation.

In 2016, the Centers for Disease Control and Prevention reported that SCD affects approximately 100,000 Americans and costs the United States over $475 million a year in healthcare.[2] More than two million people are genetic carriers of sickle cell trait (SCT).[3] Derlega et al. (2014) stated that though other racial and ethnic groups are affected by SCD, African Americans represent the largest population in the United States with SCD.[4]

Advances in preventive care and medical therapy over the past 30 years have led to increased survival for children born with SCD, currently in excess of 90 percent.[5] However, progress has been slow. The National Institutes of Health (NIH)

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[1] Siddiqi AE, Jordan LB, Parker CS. Sickle cell disease--the American saga. Ethn Dis 2013;23:245-8.

[2] Centers for Disease Control and Prevention, U.S. National Center for Health Statistics (2016). Chronic illness definition. https://www.cdc.gov/chronicdisease/stats/

[3] Thompson, W. E., & Eriator, I. (2014). Pain control in sickle cell disease patients: use of Complementary and Alternative medicine. Pain Medicine 15(2), 241-246. doi:10.1111/pme.12292

[4] Derlega, V. J., Janda, L. H., Miranda, J., Chen, I. A., Goodman, B., & Smith, W. (2014). How patients' self-disclosure about sickle cell pain episodes to significant others relates to living with sickle cell disease. Pain Medicine 15(9), 1496-1507.doi:10.1111/pme.12535

[5] Quinn CT, Rogers ZR, Buchanan GR. Survival of children with sickle cell disease. Blood 2004;103:4023-7.

recommended newborn screening for SCD in 1987, but it was not recognized in all 50 states and the District of Columbia until 2006. Texas was one of the initial states to start screening in 1983. The most promising disease modifying therapy for SCD, hydroxyurea,[6] was approved by the Federal Drug Administration (FDA) for adults in 1998 but did not receive approval for use in children until 2017.

After the approval of hydroxyurea, the SCD community did not see another medication become available until 2017, a gap of 19 years. Since then, other therapies have been FDA approved (Appendix C). There are discrepancies in both NIH and private research funding for SCD when compared to other rarer inherited diseases such as cystic fibrosis (CF). For instance, in 2006 it was first reported that funding for SCD worked out to be about $1,130 per person with SCD, but CF funding was $9,340 per person with CF. This difference has continued to widen. In 2020, funding for SCD is at about $914 per person with SCD, and funding for CF is at $10,497 per person with CF. Furthermore, CF patients have had three times more medical therapies approved than SCD patients during this time period.[7]

In response to the significant disparities experienced by individuals with SCD in research funding, treatment options, and access to care, the American Society of Hematology (ASH) released a SCD Report Card in 2016 to highlight these issues. Key findings from the ASH SCD Report Card include the following:

- Of adults with SCD, 75 percent did not receive hydroxyurea,
- Only one-third of children with SCD received appropriate stroke screening, and
- Mortality has improved very little for adults with SCD over the past 30 years.[8]

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[6] Segal JB, Strouse JJ, Beach MC, et al. Hydroxyurea for the treatment of sickle cell disease. Evid Rep Technol Assess (Full Rep) 2008:1-95.

[7] Farooq F, Mogayzel PJ, Lanzkron S, Haywood C, Strouse JJ. Comparison of US Federal and Foundation Funding of Research for Sickle Cell Disease and Cystic Fibrosis and Factors Associated with Research Productivity. JAMA Netw Open 2020;3:e201737.

[8] American Society of Hematology, et al. (2016). State of Sickle Cell Disease: 2016 Report. http://www.scdcoalition.org/pdfs/ASH%20State%20of%20Sickle%20Cell%20Disease%202016%20Report.pdf

Though a study from 1979 to 2005 found that the mortality rate for children with SCD decreased by three percent each year, the mortality rate for adults with SCD increased by one percent each year during the same period.[8] SCD is also associated with high treatment costs. 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.[8] There are many barriers to quality care, including health insurance, provider experience and knowledge, provider shortages, geography, economic status, and co-existing conditions.[9] Children with SCD generally have more access to care than adults with SCD, but they do not get the antibiotics they need to prevent complications.[9] Some patients are unable to fill their pain relief prescriptions, and implicit bias remains a barrier to quality care.[9]

The transition to adulthood is also problematic. Many adults are forced to rely on urgent care, leading to increased health care costs and the potential for inadequate or inappropriate treatment. [5]

The ASH SCD Report Card was updated in 2018 and reported over 40 new therapies in the research pipeline as well as the development of programs to train community health workers with comprehensive knowledge of SCD and the formation of the Sickle Cell Disease Coalition.[9]

Public awareness, attention, treatment, and accountability are essential in understanding the lives of individuals living with SCD/SCT. SCD is complex and has a vast impact on the quality of life for the individual as well as their family. Effective communication and advocacy for individuals living with SCD/SCT include access to ongoing comprehensive healthcare, integrity, empathy, implementation of recommendations, and public awareness. Public awareness of SCD/SCT requires the attention of the community and health care workers, as well as local, state, and federal agencies. These entities must understand and work in unity to help solve healthcare disparities involving individuals with SCD/SCT.

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[9] American Society of Hematology, et al. (2018). State of Sickle Cell Disease: 2018 Report. http://www.scdcoalition.org/2018%20SCD%20Report%20Card.pdf

The first Sickle Cell Advisory Committee (Advisory Committee) for the state began in 2016. The Advisory Committee, over the course of two years, developed a set of recommendations (Appendix D) for the Texas Legislature to increase awareness of SCD/SCT as well as establish a Sickle Cell Task Force (Task Force) to continue the work of the Advisory Committee. Recommendations and strategies from the Task Force will help alleviate the lack of public awareness, accountability, healthcare access, timely and appropriate treatment, and general understanding of the disease among individuals living with SCD/SCT and their families.

The Task Force is made up of seven members appointed by the Health and Human Services Executive Commissioner. Membership includes physicians specializing in hematology, members of community-based organizations who serve those with SCD, members of the public who have SCD or SCT or are the parent of a child with SCD or SCT, and a representative of a health-related institution. See Appendix A for a list of Task Force members.

3. Task Force Work Summary – Fiscal Year 2020

As required by statute, the following summarizes the Sickle Cell Task Force's (Task Force) work. The Task Force was established by the 86th Legislature in 2019, and, as such, this is the Task Force's first legislative report. Much of work of the Task Force in its first year focused on developing necessary administrative mechanisms and planning for future work.

Task Force Actions (2019-2020)

During their first year of operation, the Sickle Cell Task Force (Task Force) held four meetings (Appendix A). First-year accomplishments include the appointment of a presiding officer, Chair Dr. Michelle Noble Mackey, and the approval of Task Force bylaws. Meeting minutes and bylaws can be reviewed on the Task Force's webpage dshs.texas.gov/newborn/committees/SCTF-Business.aspx.

The Task Force also met with subject-matter experts regarding the 2018 Sickle Cell Advisory Committee (Advisory Committee) recommendations. This led to the development of four subcommittees - Milestone Development, Public Awareness Campaigns, Medicaid Contracts, and Legislatively Mandated Report - to further address the recommendations and responsibilities of the Task Force.

The Milestone Development subcommittee crafted a set of milestones to address the 2018 Advisory Committee recommendations. These milestones are discussed in the next section of this report.

Task Force productivity was significantly impacted by the COVID-19 pandemic, including the cancellation of one meeting and the inability to hold in-person meetings.

Future Activity

Based on information provided by subject-matter experts and discussions within the Task Force, the following represents the plan for future work.

Milestones for Developing Public Awareness Campaigns

The Task Force worked with DSHS to develop a short-term awareness campaign in Texas for September 2020, which is nationally-designated as Sickle Cell Awareness month. This campaign included information regarding COVID-19 as patients with sickle cell disease (SCD) are vulnerable to more severe complications from COVID-19 and to the changes in medical access during the pandemic.

Long-term, the Task Force plans to utilize online messaging platforms to increase public awareness, specifically to address the genetics behind SCD inheritance and provide education about SCD symptoms and treatments. Emphasis will also be placed on how SCD/sickle cell trait (SCT) can impact any individual, which will help destigmatize and demystify the notion that SCD/SCT only affects people who are Black or of African descent. It will also be important to communicate the estimated population in Texas affected by SCD/SCT using data from the Newborn Screening Program.

Due to present and past social injustices towards SCD/SCT patients and disparities in health care quality and access, the Task Force also intends to develop a focused campaign on health care education targeting health care professionals and hospitals. The Task Force plans to engage in a collaborative approach that focuses on community-based organizations that provide education to the community as well as seek ongoing input from experts. Moreover, the Task Force plans for the campaign to address the relative lack of access to treatment and shortage of physicians who are knowledgeable about SCD.

Milestones for Studying Sickle Cell Surveillance

The Task Force plans to review current surveillance mechanisms in the state and invite representative(s) from the Texas Syndromic Surveillance System to assess how to incorporate surveillance of SCD. The Task Force will explore funding and mentorship opportunities for state-led surveillance programs. The Task Force will also review current birth reporting processes to identify areas of improvement, including hospital and physician challenges. Long-term, the Task Force will plan to review state record-keeping requirements for data collection and storage and recommend new protocols or changes to protocols for surveillance.

Milestones for partnering with Medicaid/Medicare, Managed Care and Accountable Care Organizations

The Task Force will review current managed care organization (MCO) contracts for opportunities to improve healthcare for SCD patients. Further, the Task Force will

collaborate with MCO representatives to seek ways to improve access points to comprehensive SCD-focused health care (e.g. Sickle Cell medical homes), coverage of novel medical therapies, and adherence to preventive care and monitoring, such as immunizations and stroke screening. Long-term, the Task Force will review, make further recommendations, or both, for optimizing the relationship between MCOs and SCD patients.

Milestones for Working with Community Health Workers

The Task Force will review educational modules by DSHS that provide SCD/SCT education and recommend areas for improvement. The Task Force will seek to understand locations where individuals with SCD seek care, including Urgent Care Clinics, for possible targeted educational efforts. Furthermore, the Task Force will review community health worker (CHW) roles and responsibilities and make recommendations to optimize their use in improving the care of patients with SCD in Texas. The Task Force also plans to develop a lecture or conference for CHWs in the state to provide education on sickle cell, available resources, and discuss opportunities for CHW optimization.

Task Force Costs

The costs associated with the Task Force were for DSHS administrative support related to meeting preparation, planning, and follow-up. Members of the Task Force did not receive reimbursement for travel expenses to Task Force Meetings.

4. Conclusion

The Sickle Cell Task Force (Task Force), with administrative support from Department of State Health Services staff, has prioritized its discussions around the Task Force mandate to identify strategies to raise awareness of Sickle Cell Disease and Sickle Cell Trait and reviewing and advising the department on implementing the recommendations made in the 2018 Sickle Cell Advisory Committee Report published by the Sickle Cell Advisory Committee. In its first year of operation, the Task Force has appointed a presiding officer, formed subcommittees, and set milestones to address the recommendations made in the 2018 report. The Task Force will continue its work to meet these milestones.

List of Acronyms

Acronym	Full Name
ASH	American Society of Hematology
CF	Cystic Fibrosis
CHW	Community Health Worker
DSHS	Department of State Health Services
FDA	Federal Drug Administration
MCO	Managed Care Organization
NIH	National Institutes of Health
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait

Appendix A. Task Force Members and Attendance

Table 1. Sickle Cell Task Force Members

Member	Position/Category
Dr. Titilope Fasipe	Representative of a health-related institution
Dr. Melissa Frei-Jones	Physician specializing in hematology
Dr. Michelle N. Mackey, Chair	Member of the public who has sickle cell disease or is the parent of a person with sickle cell disease or trait
Dr. Alecia Nero	Physician specializing in hematology
Ms. Tonya Prince	Member from a community-based organization with experience addressing the needs of individuals with sickle cell disease
Ms. Marqué Reed-Shackelford	Member of the public who has sickle cell disease or is a parent of a person with sickle cell disease or trait
Ms. Alysian Thomas, J.D.	Member from a community-based organization with experience addressing the needs of individuals with sickle cell disease

Table 2. Task Force Meeting Dates and Attendance during the First Year

Date	Attendance	Quorum
November 26, 2019	Seven of the seven members participated, six via teleconference and one in person.	Yes
February 27, 2020	Seven of the seven members participated in person in Austin, Texas.	Yes
June 12, 2020	Seven of the seven members participated via teleconference.	Yes
August 31, 2020	Seven of the seven members participated via teleconference.	Yes

Appendix B. Timeline of Sickle Cell Legislation

Table 3. Timeline of National Sickle Cell Legislation

Year	Legislation	Description
1972	National Sickle Cell Anemia Control Act	Authorized funding for research, screening and counseling programs and development and dissemination of educational materials. President Richard Nixon pledged that his administration would "reverse the record of neglect of the dreaded disease."[i]
1983	National Sickle Cell Awareness Month	Recognition of national Sickle Cell Awareness month was first introduced in a resolution by the Congressional Black Caucus.[ii] President Ronald Reagan issued Proclamation 5102 to recognize September 1983 as National Sickle-Cell Anemia Awareness Month.[iii]
2004	Sickle Cell Treatment Act of 2003	Included in the American Jobs Creation Act signed into law in 2004.[iv] Amends the Social Security Act to include an Optional Medicaid benefit primary and secondary medical strategies, treatment, and services for individuals living with SCD. Includes Federal reimbursement to States 50% of costs for services to identify and educate eligible Medicaid recipients who have SCD or who are carriers of the sickle cell gene, and for education regarding the risks of stroke and other medical complications. Directs the Administrator of the Health Resources and Services Administration to: (1) conduct a demonstration program by making grants to up to 40 federally-qualified and nonprofit health care providers for the development and establishment of systemic mechanisms to improve the prevention and treatment of sickle cell disease; and (2) contract with an entity to serve as the National Coordinating Center for the demonstration program. Sets forth requirements for awarding grants to establish the demonstration program. Authorizes appropriations for FY 2005 through 2009.[v]

Year	Legislation	Description
2018	Sickle Cell Disease and Heritable Blood Disorders Research, Surveillance, Prevention and Treatment Act of 2018	Signed into law on December 18, 2018. Reauthorizes a sickle cell disease grant program for federally-qualified health centers, nonprofit hospitals or clinics, and university health centers through 2023. Also provides the authority to award grants to government, educational, and nonprofit entities for the purpose of supporting data collection and public health activities regarding heritable blood disorders, including SCD.[vi]
2020	National Sickle Cell Disease Awareness Month	President Trump issued a proclamation designating September 2020 as National Sickle Cell Disease Awareness month. Stated the goal of extending lives of Americans with SCD by 10 years and finding a cure by 2029.[vii]

[i] Nixon, Richard. Statement on Signing the National Sickle Cell Anemia Control Act. Online by Gerhard Peters and John T. Woolley, The American Presidency Project. https://www.presidency.ucsb.edu/node/254792

[ii] Hartsell, Malerie. (2017, September 18). National Sickle Cell Awareness Month. South Carolina Department of Health and Environmental Control. https://blog.scdhec.gov/2017/09/18/national-sickle-cell-awareness-month-2/

[iii] Reagan, Ronald. (1983, September 21). Proclamation 5102 – National Sickle-Cell Anemia Awareness Month, 1983. https://www.reaganlibrary.gov/research/speeches/92183i

[iv] Davis, Danny K. (2018, November 29). Sickle Cell Renewal Passes! https://davis.house.gov/statements/sickle-cell-renewal-passes/

[v] Congressional Research Service. (2004, October 22). Summary: H.R.4520 – American Jobs Creation Act of 2004. https://www.congress.gov/bill/108th-congress/house-bill/4520

[vi] Congressional Research Service. (2018, December 18). Summary: S.2465 – Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention, and Treatment Act of 2018. https://www.congress.gov/bill/115th-congress/senate-bill/2465

[vii] Trump, Donald. (2020, August 31). Proclamation on National Sickle Cell Disease Awareness Month, 2020. https://www.whitehouse.gov/presidential-actions/proclamation-national-sickle-cell-disease-awareness-month-2020/

Appendix C. FDA-Approved Sickle Cell Therapies

Table 4. Therapies approved by the FDA for treatment of SCD

Date Approved	Drug	Description
1998	Hydroxyurea	Approved for adults only to reduce complications.[i]
July 2017	Endari (L-glutamine oral powder)	First new treatment in almost 20 years. Approved for patients ages 5 and older to reduce complications of sickle cell disease.[ii]
December 2017	Hydroxyurea	Approved for pediatric patients ages 2 and older with sickle cell anemia to reduce the frequency of painful crises and need for blood transfusions.[iii]
November 2019	Adakveo (Crizanlizumab-tmca)	Approved for patients 16 years and older to reduce the frequency of vaso-occlusive crisis. The first targeted therapy approved for SCD.[iv]
November 2019	Oxbryta (voxelotor)	Approved for patients ages 12 and older. Affects red blood cells, making it less likely for sickle cells to bind together.[v]

[i] Federal Drug Administration. (2018, June 18). The FDA Encourages New Treatments for Sickle Cell Disease. https://www.fda.gov/consumers/consumer-updates/fda-encourages-new-treatments-sickle-cell-disease.

[ii] Trump, Donald. (2020, August 31). Proclamation on National Sickle Cell Disease Awareness Month, 2020. https://www.whitehouse.gov/presidential-actions/proclamation-national-sickle-cell-disease-awareness-month-2020/

[iii] Federal Drug Administration. (2017, December 21). FDA approves hydroxyurea for treatment of pediatric patients with sickle cell anemia. https://www.fda.gov/drugs/resources-information-approved-drugs/fda-approves-hydroxyurea-treatment-pediatric-patients-sickle-cell-anemia#:~:text=On%20December%2021%2C%202017%2C%20the,moderate%20to%20severe%20painful%20crises.

[iv] Federal Drug Administration. (2019, November 15). FDA approves first targeted therapy to treat patients with painful complication of sickle cell disease. https://www.fda.gov/news-events/press-announcements/fda-approves-first-targeted-therapy-treat-patients-painful-complication-sickle-cell-disease

[v] Federal Drug Administration. (2019, November 25). FDA approves novel treatment to target abnormality in sickle cell disease. https://www.fda.gov/news-events/press-announcements/fda-approves-novel-treatment-target-abnormality-sickle-cell-disease

Appendix D. Recommendations from the 2018 Sickle Cell Advisory Committee Annual Report

Recommendation 1: Establish a Sickle Cell Task Force

Due to the abolishment of the Sickle Cell Advisory Committee, the Department of State Health Services (DSHS) added two new member positions to the Newborn Screening Advisory Committee (NBSAC) to focus on SCD and SCT. However, the NBSAC oversees a total of 53 conditions. A task force solely focused on SCD and SCT would be able to make further progress on these conditions than if included within the NBSAC's wider scope.

As such, the Committee recommends that DSHS create a task force to continue the work of the Sickle Cell Advisory Committee. The task force should have broader objectives than those of the Committee and focus on increasing stakeholder involvement and improving the efficiency of outreach efforts. The Committee also recommends that funding be provided to support the work and objectives of the task force.

Recommendation 2: Develop Statewide Sickle Cell Awareness Campaigns

The Committee recommends that the state allocate funds to develop SCD/SCT awareness campaigns. Such campaigns should reach a broad audience with specific and actionable messages, and engage state medical boards, professional healthcare organizations, and community-based organizations. The Committee also recommends that the use of the Texas Health Steps Sickle Cell Module as an educational resource should be also be included in the awareness campaigns. These statewide campaigns should involve a university-based marketing team or professional marketing firm.

Recommendation 3: Begin Statewide Sickle Cell Surveillance Throughout the Lifespan

The Committee recommends that DSHS assess the existing state surveillance system framework to determine an appropriate method of collecting data on SCD and SCT. Any future SCD/SCT surveillance should identify and monitor the critical period of transition from pediatric care to adult care due to increased morbidity and mortality during this period.

DSHS should also contribute to the national conversation on SCT and SCD by sharing Texas surveillance statistics with national organizations such as the United States Centers for Disease Control and Prevention (CDC), the Sickle Cell Disease Association of America, and the Sickle Cell Disease Coalition.

Recommendation 4: Partner with Medicaid/Medicare, Managed Care Organizations (MCO), and Accountable Care Organizations (ACO)

The Committee recommends that HHSC Texas Medicaid expand Medicaid contracts with ACOs and MCOs to ensure the availability of services and raise awareness amongst healthcare professionals to provide the full array of Medicaid and Medicare benefits. HHSC Texas Medicaid should also promote the use of the medical home model and case management services to raise awareness and improve access for people with SCD, including expansion of covered medical services for adults.

Recommendation 5: Utilize Community Health Workers to Improve Care for those with SCD and SCT

The Committee recommends the use of CHWs to help improve patient care, navigation, education, and the transition from pediatric to adult services at the individual and community level through outreach and support. DSHS should also require sickle cell disease education as part of the CHW certification.