2018 Sickle Cell Advisory Committee Final Report

As Required by

Texas Government Code,

Section 531.012

25 Texas Administrative Code,

Section 37.420

Sickle Cell Advisory Committee

December 2018

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Executive Summary

In June 2016, under the authority of Texas Government Code, Section 531.012, Health and Human Services adopted 25 Texas Administrative Code, Section 37.420 to establish the Department of State Health Services (DSHS) Sickle Cell Advisory Committee (Committee). The Committee is charged with reviewing strategies for and making recommendations to raise public awareness of sickle cell disease (SCD) and sickle cell trait (SCT) in Texas.

The following are the major activities of the Committee during fiscal years 2017-2018:

- development of accessible educational resources for medical professionals, caregivers, and patients with SCD;
- recommendation of strategies to raise public awareness of SCT and SCD to the Executive Commissioner of the Health and Human Services Commission (HHSC);
- creation of the Sickle Cell Resources webpage; and
- update of the DSHS Newborn Screening Website.

In 2018, the Committee finalized the following recommendations to raise awareness of SCD and SCT in Texas:

- establish a sickle cell task force;
- develop statewide sickle cell awareness campaigns;
- begin statewide sickle cell surveillance throughout the lifespan;
- partner with Medicaid/Medicare, Managed Care Organizations, and Accountable Care Organizations to improve awareness of SCD and SCT and access to services; and
- utilize community health workers to improve patient care.

The Committee was abolished on September 1, 2018, in accordance with 25 Texas Administrative Code, Section 37.420. In an effort to continue the work of the Committee, DSHS added two new member positions representing the sickle cell community on the Newborn Screening Advisory Committee.

1. Introduction

In June 2016, under the authority of Texas Government Code, Section 531.012, the Sickle Cell Advisory Committee (Committee) was established by 25 Texas Administrative Code, Section 37.420 to raise public awareness of sickle cell disease (SCD) and sickle cell trait (SCT) in Texas with the following responsibilities:

- review and suggest ways to raise public awareness of sickle cell disease and sickle cell trait; and
- recommend two specific strategies to raise public awareness by the end of fiscal year 2018.

The Committee must submit an annual report to the Texas Health and Human Services Commission (HHSC) Executive Commissioner on or before December 1, 2018. The Committee must also report any recommendations to the HHSC Executive Commissioner at a meeting of the HHSC Executive Council established under Texas Government Code, Section 531.0051. Additionally, the Committee must submit an annual written report to the Texas Legislature of any policy recommendations made to the HHSC Executive Commissioner.

In accordance with 25 Administrative Code, Section 37.420, the report outlines the following:

- a summary of the Committee's outcomes during their first year of operation;
- a description of Committee meetings, attendance (Appendix A), activities, and costs during their second year; and
- recommendations proposed by the committee to the HHSC Executive Commissioner to raise awareness of SCD and SCT.

2. Background

Sickle Cell Disease (SCD) affects at least 100,000 individuals in the United States.[1] SCD results from a gene mutation that causes red blood cells (RBCs) to become fragile, breakdown rapidly, and change shape from a round disc to a *sickle* shape. Individuals who inherit one copy of the sickle cell gene have sickle cell trait (SCT) and rarely show symptoms. Those who inherit two copies of the gene mutation have the most common form of SCD called Homozygous SS Disease. Anemia, acute and chronic pain, and damage to multiple organ systems including the brain, lungs, kidneys, eyes, and spleen may result when sickle-shaped RBCs cannot move easily through the blood vessels. During a lifetime, SCD is marked by unpredictable episodes of excruciating pain resulting in frequent school and work absence disrupting education, employment, and social relationships.

All babies born in Texas are screened for a number of inherited disorders through the DSHS newborn bloodspot screening program, including SCD and SCT. In Texas, approximately 180 children each year are diagnosed with SCD (Appendix B) and over 5,000 children are identified with SCT annually (Appendix C). Through early identification and intervention, infants with SCD benefit from preventive care, including referrals to hematology specialists and prophylactic antibiotics to prevent life-threatening infections and death from sepsis. As a result, children with SCD are living longer with more than 90 percent expected to survive to adulthood.[2]

However, during the transition from pediatric care to adult health care services, young adults with SCD experience increased morbidity and mortality due to lack of access to care, including accessibility to medical services and insurance coverage.[2] Awareness of the condition among the general public is also deficient. During a lifetime, a person with SCD will interact with primary care providers, school nurses, teachers, athletic coaches, emergency departments, insurance carriers, case managers, pharmacists, admissions counselors, professors, university health centers, and employers. Overall knowledge of this blood disorder among these

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[1] Brousseau DC, Panepinto JA, Nimmer M, Hoffmann RG. The number of people with sickle-cell disease in the United States: national and state estimates. *Am J Hematol.*85(1):77-78.

[2] Quinn CT, Rogers ZR, McCavit TL, Buchanan GR. Improved survival of children and adolescents with sickle cell disease. *Blood.*115(17):3447-3452.

individuals is poor and is a barrier to care for people with SCD. [3,4] This may ultimately result in a lack of recognition of the severity of SCD symptoms or a lack of appropriate accommodations to manage risk.

In addition, due to potential exercise-induced complications associated with SCT, coaches, athletes, trainers, and athletic programs need to be informed of the risk to students and athletes. Possible exercise and dehydration induced complications in people with sickle cell trait include hematuria, rhabdomyolysis, and increased susceptibility to heat stroke. Precautions should be taken, such as modifying exercises, to prevent severe complications.

There is a need to increase public awareness of SCT as part of reproductive counseling. SCT is a reproductive risk factor for SCD and may be associated with a few rare, but life-threatening complications. Though SCT and SCD can affect people of every race and ethnicity, rates among Black Americans are particularly high, wherein 1 in 12 individuals are SCT carriers. [5] In the United States, most people are unaware of their sickle cell status or the status of their reproductive partner.[5] When two individuals who both carry SCT have a child, there is a 25 percent chance that the child will have SCD, 50 percent chance that the child will have SCT, and 25 percent chance that the child will have normal hemoglobin.

The Sickle Cell Advisory Committee (Committee) was made up of seven members appointed by the HHSC Executive Commissioner and was required to meet three times per year. Membership included physicians specializing in hematology, members of community-based organizations who serve those with SCD, and individuals directly affected by SCD and SCT. Refer to Appendix A for a list of Committee members.

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[3] Whiteman LN, Haywood C, Jr., Lanzkron S, Strouse JJ, Feldman L, Stewart RW. Primary Care Providers' Comfort Levels in Caring for Patients with Sickle Cell Disease. *South Med J.* 2015;108(9):531-536.

[4] King AA, Tang S, Ferguson KL, DeBaun MR. An education program to increase teacher knowledge about sickle cell disease. *J Sch Health.* 2005;75(1):11-14.

[5] Boyd JH, Watkins AR, Price CL, Fleming F, DeBaun MR. Inadequate community knowledge about sickle cell disease among African-American women. *J Natl Med Assoc.* 2005;97(1):62-67.

3. Committee Activities – Fiscal Years 2017 and 2018

Committee Actions and Outcomes

First Year (2016-2017)

During the first year, the Sickle Cell Advisory Committee (Committee) held four meetings (Appendix A). First-year accomplishments included updates to the Department of State Health Services (DSHS) Newborn Screening Website and the creation of a Sickle Cell Resources webpage. All meeting minutes can be found on the Committee webpage at dshs.texas.gov/newborn/SCACbusiness.aspx.

The Committee's 2017 annual report to the Health and Human Services Commission's (HHSC) Executive Commissioner can be viewed at dshs.texas.gov/legislative/2017-Reports/Sickle-Cell-Advisory-Committee-Report.pdf.

Second Year (2017-2018)

In the second year of operation, the Committee held three meetings (Appendix A). These meetings consisted of presentations on specific topics related to sickle cell disease (SCD) and sickle cell trait (SCT) awareness, planning activities, and discussing potential recommendations to the HHSC Executive Commissioner. Official Committee meeting minutes can be found at dshs.texas.gov/newborn/SCACbusiness.aspx.

To develop informed recommendations to submit to the HHSC Executive Commissioner, the Committee sought presentations and discussion on the following topics.

- Best practices in strategic planning to improve health care coordination.
- The different types of community health workers (CHWs), the process of training CHWs, and how they potentially could be utilized to raise awareness of SCD and SCT.
- Potential marketing strategies to share information about SCD and SCT with health care professionals and communities.
- Current projects at the United States Centers for Disease Control and Prevention (CDC) that focus on sickle cell surveillance.
- The Texas legislative process and how to complete their legislative report.

- Medicaid and Children's Health Insurance Program (CHIP) health care policies and sickle cell awareness among health care professionals.

As a result of these meetings and interim work performed, the Committee was able to review and update information on the DSHS SCD and SCT webpages, as well as develop recommendations for the HHSC Executive Commissioner.

Future Activity

The Committee was abolished on September 1, 2018, in accordance with 25 Texas Administrative Code, Section 37.420. In an effort to continue the work of the Committee, DSHS added two new member positions to the Newborn Screening Advisory Committee (NBSAC) representing the sickle cell community.

Committee Costs

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

4. Committee Recommendations

The Sickle Cell Advisory Committee (Committee) proposes the following recommendations to the Health and Human Services Commission (HHSC) Executive Commissioner to raise public awareness of sickle cell disease (SCD) and sickle cell trait (SCT) in Texas.

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.

5. Conclusion

This report describes meetings activities, member attendance, actions, accomplishments, and costs of the Committee during its two-year term. Meetings involved presentations, planning, and discussions around the Committee mandate to identify strategies to raise awareness of SCD and SCT. In two years of operation, the Committee developed and updated the online educational resources on SCD and SCT.

Through these activities, and with administrative support from DSHS, the Committee identified existing resources in Texas and other states in considering the strategies to raise public awareness of SCD and SCT. The Committee developed five recommendations for the HHSC Executive Commissioner

- establish a Sickle Cell Task Force;
- develop statewide sickle cell awareness campaigns;
- begin statewide sickle cell surveillance throughout the lifespan;
- partner with Medicaid/Medicare, Managed Care Organizations and Accountable Care Organizations to improve awareness of SCD and SCT and access to services; and
- utilize community health workers to improve care for those with SCD and SCT.

The Committee is looking for support, funding, and implementation of these strategies by HHSC and encourages taking action on the recommendations to continue impacting the lives of thousands of Texans with SCD and SCT.

List of Acronyms

Acronym	Full Name
ACO	Accountable Care Organization
CDC	United States Centers for Disease Control and Prevention
CHW	Community Health Worker
CHIP	Children's Health Insurance Program
DSHS	Department of State Health Services
HHSC	Health and Human Services Commission
HSR	Health Service Region
MCO	Managed Care Organization
NBSAC	Newborn Screening Advisory Committee
OOS	Out of State
RBC	Red Blood Cells
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait

Appendix A. Committee Members and Attendance

Table 1. Sickle Cell Advisory Committee Members

Member	Email	Position/Category
Dr. Titilope Fasipe	titilope.ishola@gmail.com	Member of a Community Based Organization
Ms. Alysian Thomas	alysianthomas@yahoo.com	Member of a Community Based Organization
Dr. Melissa Frei-Jones	freijones@uthscsa.edu	Physician Specializing in Hematology
Dr. Clarissa Johnson	Clarissa.Johnson@cookchildrens.org	Physician Specializing in Hematology
Dr. Harinder Juneja	junerano@aol.com	Health Professional in an Academic Setting (Chair)
Ms. Toni Tennent	ttennent@gmail.com	Parent or Individual with Sickle Cell Disease or Trait
Ms. Marque Reed-Shackelford	mrshackelford23@gmail.com	Parent or Individual with Sickle Cell Disease or Trait

Table 2. Committee Meeting Dates and Attendance during the First Year

Date	Attendance	Quorum
August 26, 2016	Six of the seven members participated via teleconference.	Yes
November 4, 2016	Six of the seven members participated via teleconference.	Yes
March 24, 2017	Seven of the seven members participated in-person/Austin, TX.	Yes
July 21, 2017	Six of the seven members participated via teleconference.	Yes

Table 3. Committee Meeting Dates and Attendance during the Second Year

Date	Attendance	Quorum
November 3, 2017	Five of the seven members participated via teleconference.	Yes
March 30, 2018	Seven of the seven members participated in-person/Austin, TX.	Yes
July 27, 2018	Six of the seven members participated in-person/Austin, TX; one participated via teleconference.	Yes

Appendix B. Hemoglobin Diagnosed Cases

Table 3. Hemoglobin Diagnosed Cases 2008-2017 by Calendar Year

Year	Sickle / Beta Plus Thalassemia	Sickle / Beta Zero Thalassemia	Sickle C Disease	Sickle Cell Anemia	Total
2008	11	6	50	89	**156**
2009	16	3	59	104	**182**
2010	9	3	54	70	**136**
2011	17	2	45	83	**147**
2012	16	4	51	104	**175**
2013	21	3	56	92	**172**
2014	20	2	54	102	**178**
2015	9	2	47	117	**175**
2016	20	2	39	133	**194**
2017	17	3	45	128	**193**
Total	**156**	**30**	**500**	**1,022**	**1,708**

SOURCE: Department of State Health Services Laboratory Information Management System

Table 4. Hemoglobin Diagnosed Cases 2008-2017 by Race / Ethnicity

Race / Ethnicity	Sickle / Beta Plus Thalassemia	Sickle / Beta Zero Thalassemia	Sickle C Disease	Sickle Cell Anemia	Total	Percent of Total
AFRICAN AMERICAN	130	17	446	871	1,464	**86**
AMERICAN INDIAN	0	0	1	1	2	**0**
ASIAN	2	0	1	1	4	**0**
HISPANIC	12	6	14	54	86	**5**
UNKNOWN	3	1	9	16	29	**2**
OTHER	5	4	26	68	103	**6**
WHITE	4	2	3	11	20	**1**
Total	**156**	**30**	**500**	**1,022**	**1,708**	**100%**

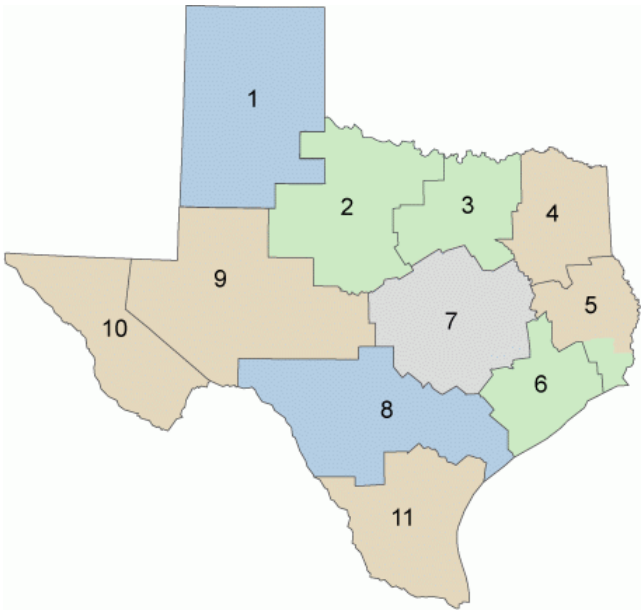
SOURCE: Department of State Health Services Laboratory Information Management System

Table 5. Hemoglobin Diagnosed Cases 2008-2017 by Health Service Region (HSR)

HSR	Sickle / Beta Plus Thalassemia	Sickle / Beta Zero Thalassemia	Sickle C Disease	Sickle Cell Anemia	Total	Percent of Total
HSR 1	3	1	7	14	25	**1**
HSR 2	1	0	3	11	15	**1**
HSR 3	40	11	188	360	599	**35**
HSR 4	9	3	22	37	71	**4**
HSR 5	7	0	19	32	58	**3**
HSR 6	68	6	190	405	669	**39**
HSR 7	22	3	38	91	154	**9**
HSR 8	5	5	16	43	69	**4**
HSR 9	0	0	6	8	14	**1**
HSR 10	0	0	0	10	10	**1**
HSR 11	0	1	2	7	10	**1**
OOS	1	0	9	4	14	**1**
Total	**156**	**30**	**500**	**1,022**	**1,708**	**100%**

SOURCE: Department of State Health Services Laboratory Information Management System

Figure 1. Texas Health Service Regions Area Map



Appendix C. Sickle Trait Cases

Table 6. Sickle Trait Cases 2008-2017 by Race/Ethnicity and Calendar Year

Year	AFRICAN AMERICAN	AMERICAN INDIAN	ASIAN	HISPANIC	OTHER	WHITE	BLANK	Total
2008	3,552	6	16	1,228	324	288	117	**5,531**
2009	3,484	4	14	1,245	317	321	123	**5,508**
2010	3,312	9	21	1,087	337	297	104	**5,166**
2011	3,234	5	17	1,048	337	301	127	**5,069**
2012	3,368	9	25	1,047	358	309	100	**5,216**
2013	3,525	8	27	1,084	372	293	128	**5,437**
2014	3,701	9	27	1,167	488	333	131	**5,856**
2015	3,970	14	34	1,130	491	323	109	**6,071**
2016	4,008	4	33	1,185	590	360	117	**6,297**
2017	3,907	13	32	1,084	579	357	116	**6,088**
Total	**36,061 (64.1%)**	**81 (.1%)**	**246 (.4%)**	**11,305 (20.1%)**	**4,192 (7.5%)**	**3,182 (5.7%)**	**1,172 (2.1%)**	**56,239 (100%)**

SOURCE: Department of State Health Services Laboratory Information Management System

Table 7. Sickle Trait Cases 2008-2017 by Health Service Region (HSR) and Calendar Year

HSR #	2008	2009	2010	2011	2012	2013	2014	2015	2016	2017	TOTAL
HSR 1	93	104	85	99	97	97	104	98	111	89	**977**
HSR 2	69	64	62	62	61	53	63	67	50	60	**611**
HSR 3	1,697	1,685	1,598	1,579	1,662	1,733	1,802	1,854	1,883	1,936	**17,429**
HSR 4	333	305	335	295	298	336	285	289	288	315	**3,079**
HSR 5	218	203	182	179	197	161	182	191	190	171	**1,874**
HSR 6	1,953	1,988	1,835	1,790	1,867	2,030	2,270	2,405	2,526	2,365	**21,029**
HSR 7	488	461	453	426	419	405	434	449	460	459	**4,454**
HSR 8	308	328	294	282	276	289	323	331	385	338	**3,154**
HSR 9	57	45	57	72	63	58	101	103	101	82	**739**
HSR 10	62	76	63	56	64	69	56	73	85	76	**680**
HSR 11	253	249	202	229	212	206	234	211	216	196	**2,208**
Unknown	93	104	85	99	97	97	2	0	2	1	**5**
TOTAL	**5,531**	**5,508**	**5,166**	**5,069**	**5,216**	**5,437**	**5,856**	**6,071**	**6,297**	**6,088**	**56,239**

SOURCE: Department of State Health Services Laboratory Information Management System