Sickle Cell Advisory Committee

As Required by

25 Texas Administrative Code §37.420

With Support from the Texas Department of State Health Services

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TEXAS
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Executive Summary

In June 2016, rules were adopted under the Texas Administrative Code, Title 25, Part 1, Chapter 37, Subchapter R (25 TAC §37.420), to establish the Texas 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).

DSHS appointed committee members and held the first meeting August 2016. During fiscal year 2017, the Committee held three more meetings. Major activities and actions of the Committee include:

- The appointment of presiding officers;
- Approval of Committee bylaws;
- Identification of potential strategies to raise public awareness of SCT and SCD; and
- The development of accessible educational resources for medical professionals, caregivers and patients with SCD.

The Committee identified, explored, updated, and expanded existing resources within the State of Texas in considering the goal of raising public awareness of SCD and SCT. In the first year, the members initiated or completed the following:

- Sickle Cell Toolkit creation;
- DSHS Newborn Screening website update;
- Identified the Texas Health Steps Sickle Cell Disease and Trait Online Training Module as a possible basis for future training.

Introduction

The Sickle Cell Advisory Committee was created to raise public awareness of sickle cell disease (SCD) and sickle cell trait (SCT) in the State of Texas. The Committee is comprised of seven members appointed by the Health and Human Services Commission (HHSC) Executive Commissioner, and is required to meet three times per year. At the end of the first year, the committee is required to submit a summary report of the committee's actions and any recommendations to the HHSC Executive Commissioner.

The Texas Administrative Code (TAC) 25 TAC §37.420 establishes the tasks, reporting requirements, membership requirements, membership qualifications, and meeting schedules for the Sickle Cell Advisory Committee. The Committee is tasked to:

- Review and suggest methods for raising public awareness of sickle cell disease and sickle cell trait; and
- By the end of fiscal year 2018, recommend two specific strategies raise public awareness.

The Committee is composed of a variety of medical professionals, members of community based organizations, and individuals directly affected by SCD and SCT, both patients and caregivers, who have a vested interest in promoting awareness of the disease and the trait in order to improve the quality of life of affected individuals. Refer to Appendix A for a list of current committee members.

The Committee has met four times since establishment in 2016. The major activities and actions of the Committee include:

- The appointment of presiding officers;
- Approval of Committee bylaws;
- Identification of potential strategies to raise public awareness of SCT and SCD; and
- The development of accessible educational resources for medical professionals, caregivers and patients with SCD.

Background

SCD affects 100,000 individuals in the United States.[1] SCD is identified at birth through newborn screening in order to identify patients with disease who will benefit from early preventive care, especially prophylactic antibiotics, to prevent life-threatening infections and death from sepsis. During a lifetime, SCD is marked by unpredictable episodes of excruciating pain resulting in frequent school and work absence, and disrupts education, employment, and social relationships.

In Texas, approximately 180 children a year are diagnosed with SCD identified through newborn screening (Appendix B). Over 5,000 children are identified with SCT annually (Appendix C). Due to early detection, children with SCD benefit from referral to sickle cell specialists and preventive care. As a result, children with SCD are living longer with survival to age 18 years exceeding 90 percent.[2] The number of adults living with SCD is increasing while awareness of the condition among the general public is deficient. During a lifetime, they 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 individuals is poor and is a barrier to care for people with SCD.[3] [4] This may result in lack of recognition of severity of symptoms or lack of appropriate accommodations.

SCT, although an asymptomatic condition, is a reproductive risk factor for SCD and may be associated with a few rare, but life-threatening complications. Among black Americans, 1 in 12 people carry SCT and yet, most are unaware of their sickle cell

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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.

[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.

status or the status of their reproductive partner.[5] Because carriers of SCT have a 25 percent chance of having offspring with SCD, there is a need to increase public awareness of SCT as part of reproductive counseling. In addition, due to exercise-induced complications associated with SCT, coaches, athletes, trainers, and athletic programs need to be informed of the true risk to student, amateur, and professional athletes, as well as understand exercise modifications to prevent these severe complications.

The creation of the Sickle Cell Advisory Committee is an important first step in raising awareness of both trait and disease in Texas as a method to improve care of affected people with SCD, raise reproductive awareness among carriers, and prevent rare complications of SCT.

[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.

Committee Activities - Year One

COMMITTEE MEETING DATES AND ATTENDANCE OF MEMBERS

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 members participated in-person/Austin, TX	Yes
July 21, 2017	Six of the seven members participated via teleconference	Yes

DESCRIPTION OF COMMITTEE ACTIONS

The first meeting of Committee members occurred on August 26, 2016, and was a preliminary meeting designed to prepare members to perform their assigned duties. During the pre-meeting, members were introduced to each other and the Department of State Health Services (DSHS) staff who would be assisting the Committee. Committee members were provided with training in responsibilities, code of conduct, ethics, quorum, conflict of interest, and open meetings. A draft of Committee bylaws and an overview of DSHS rulemaking process were also provided.

The first official Committee meeting of fiscal year 2017, was held on November 4, 2016, and began with the nomination and election of Dr. Harinder Juneja as the Chairperson and Mr. LeDarrin Taite as Vice Chairman. Minutes of the August 26, 2017 meeting were approved, as well as the Committee bylaws. The majority of the meeting discussion focused on potential strategies to raise public awareness in Texas within the financial limitations of the Committee using educational resources currently available. Actions of the Committee included a request to review the Newborn Screening Program's (NBS) ACT and FACT Sheets for health care providers related to sickle cell conditions, as well as content of the DSHS websites. In addition, the Committee planned to explore educational materials found on national websites including the Centers for Disease Control Sickle Cell Webinars and

Grand Rounds. Action items from the meeting included requesting DSHS to review what other states are doing to raise awareness and to have members bring high-quality information to share with health care professionals, patients and families, and the general public including schools, universities, legislators, and insurance companies.

The second meeting occurred on March 24, 2017, and served as the annual in-person meeting. The Committee approved the November 4, 2016 meeting minutes and focused on the action items created in the previous meeting. DSHS presented research on other states activities, but only 3 of 50 states responded, mostly describing events within major cities. The Committee requested a second inquiry be sent to the states who did not respond initially, as well as research of legislation that exists in other states relating to Sickle Cell Advisory Committees. The Committee sought to explore all options available within the current State programs that could be adapted to meet the needs of raising public awareness. An action item was created to invite representatives of the Syndromic Surveillance, Title V Programs, and the Community Health Workers (CHW) to the next meeting to explore the possibility of using their platforms as a resource to piggyback off of in order to meet the goal of raising awareness of SCD and SCT. A review of the DSHS NBS program website and Sickle Cell webpages was recommended for possible updates to the format and links. The final Committee action was to initiate the creation of a Sickle Cell Toolkit, as well as explore using other media platforms, including Twitter, to raise awareness.

The third and the final required meeting for fiscal year 2017 was held on July 21, 2017. The meeting minutes from March 4, 2017 were approved. A representative from DSHS' Regional and Local Health Services gave a presentation on the Texas Syndromic Surveillance System. Syndromic surveillance allows for early detection of abnormal disease patterns that could result in high morbidity and mortality. The Committee was interested in this topic as a source for identifying and tracking SDC patients demographically, in an effort to focus public awareness and education interventions. A representative of the CHW provided background information and the Committee discussed educating CHWs in SCD and SCT as a possible means to raise awareness. The Committee suggested a review of the Sickle Cell Disease and Trait training module provided by Texas Health Steps to determine how many people and what type of medical providers access the training. Committee member, Dr. Titilope Fasipe, presented recommended updates and editing of the DSHS NBS website for SCD and SCT, followed by Committee member, Ms. Toni Tennent, who reviewed the Sickle Cell Toolkit, which includes educational information aimed at the medical community, patients, caregivers, and community in general. A suggestion was made to DSHS staff to develop bilingual materials on the website in

Spanish, as well as identify the number of CHW and their regional locations to target training. The Committee Chairperson, Dr. Juneja, agreed to begin discussions with other professionals on potential marketing strategies to review at the next meeting. The Committee created a work group to draft the annual report for review at the next meeting.

In late August 2017, DSHS was notified that the Vice Chair of the Sickle Cell Advisory Committee, Mr. LeDarrin Taite, had passed away unexpectedly leaving a vacancy on the Committee.

COMMITTEE ACCOMPLISHMENTS

A review of the NBS website and Sickle Cell webpages was recommended with updates to the format and links. Spanish translation of these documents, once finalized, was also suggested.

The Committee identified the need for educational tools which will serve as a resource for professionals, patients, and the general public. In the first committee year, the members initiated or completed the following:

- Sickle Cell Toolkit Creation;
- DSHS Newborn Screening Website update;
- Identified the Texas Health Steps Sickle Cell Disease and Trait Online Training Module as a possible basis for future training.

An extension of the term of the Committee is needed, as efforts to promote awareness of SCD and SCT need to be an ongoing process in order to reach a larger audience. Also, repetition of the message is essential for success of messaging to the target audience. An active Sickle Cell Advisory Committee helps the DSHS Commissioner monitor the progress and success of an awareness program, and possibly recommend changes in the state's approach to the endeavor. However, the Committee is not considering this as one of the two formal recommendations to the Executive Commissioner at this time.

ANTICIPATED ACTIVITIES OF COMMITTEE FOR NEXT YEAR

Activities the Committee anticipates to complete over the next year include:

- Nomination and election of a new Vice Chair for the Committee;
- Work with DSHS to solicit and appoint new member for vacant position;

- Establish a method to disseminate information developed during initial committee term year to meet goal of raising public awareness of SCD and SCT; and
- Determine the final two strategies to raise public awareness to recommend to the Executive Commissioner.

COMMITTEE COSTS

The only costs associated with the committee were for DSHS administrative support related to meeting preparation, planning, and meeting follow-up support.

Conclusion

The Sickle Cell Advisory Committee with support from DSHS staff identified existing resources in Texas and other states in considering the goal of raising public awareness of SCD and SCT during their first committee term. These resources, including websites and state services, were explored, updated, and expanded resulting in the creation of a Sickle Cell Toolkit which, contains key information to educate the medical community, patients, caregivers, and public in general. The Committee will be tasked during the second year with finalizing the recommendations to the HHSC Executive Commissioner and creating a method to disseminate the information developed, which will meet the final goal of raising public awareness of SCD and SCT.

List of Acronyms

Acronym	Full Name
CHW	Community Health Workers
DSHS	Department of State Health Services
HHSC	Health and Human Services Commission
NBS	Newborn Screening Program
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait
TAC	Texas Administrative Code

Appendix A. 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	Individual with Sickle Cell Disease or Trait
Vacant (as of 08/28/2017)		Individual with Sickle Cell Disease or Trait (Vice-Chair)

Appendix B. Hemoglobin Diagnosed Cases

Table 1. Hemoglobin Diagnosed Cases 2008-2016 by Calendar Year

Year	HGB - S Beta Plus Thalassemia	HGB - S Beta Zero Thalassemia	HGB - SC Sickle C Disease	HGB - SS Sickle Cell Anemia	Total
2008	10	6	45	74	**135**
2009	15	3	51	90	**159**
2010	8	3	50	69	**130**
2011	17	2	43	79	**141**
2012	15	4	51	100	**170**
2013	21	3	54	90	**168**
2014	20	2	54	103	**179**
2015	9	2	47	117	**175**
2016	18	2	39	129	**188**
Total	**133**	**27**	**434**	**851**	**1,445**

Source: Department of State Health Services Laboratory Information Management System

Table 2. Hemoglobin Diagnosed Cases 2008-2016 by Race/Ethnicity

Race/Ethnicity	HGB - S Beta Plus Thalassemia	HGB - S Beta Zero Thalassemia	HGB - SC Sickle C Disease	HGB - SS Sickle Cell Anemia	Total	Percent of Total
AFRICAN AMERICAN	114	15	385	727	**1,241**	86%
AMERICAN INDIAN			1	1	**2**	0%
ASIAN	2		1	1	**4**	0%
HISPANIC	9	6	13	44	**72**	5%
Unknown		1	9	13	**23**	2%
OTHER	4	3	23	60	**90**	6%
WHITE	4	2	2	5	**13**	1%
Total	**133**	**27**	**434**	**851**	**1,445**	100%

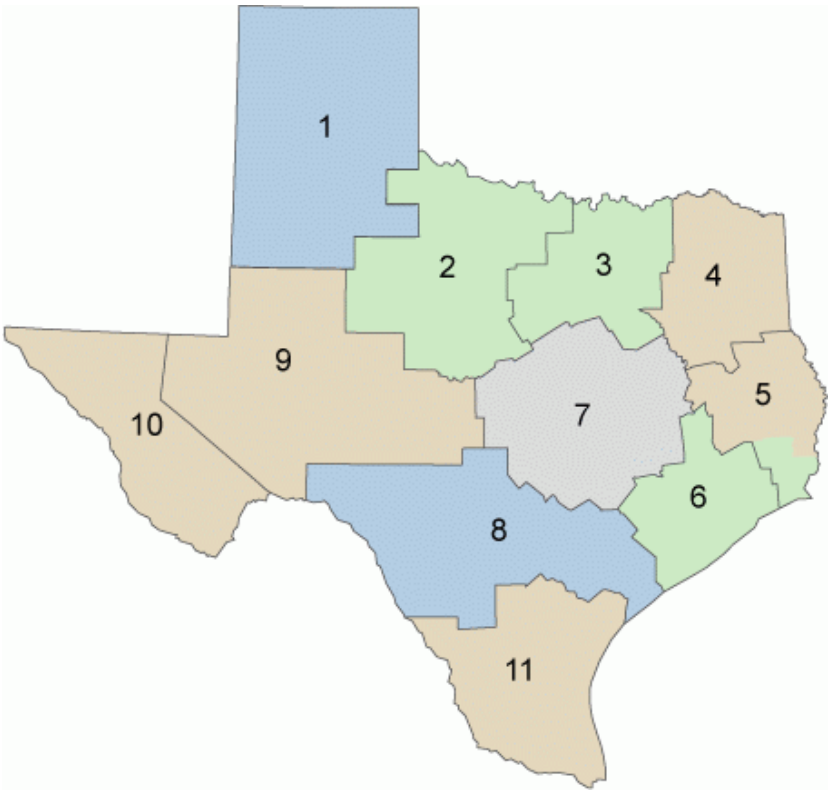
Source: Department of State Health Services Laboratory Information Management System

Table 3. Hemoglobin Diagnosed Cases 2008-2016 by Public Health Region (PHR)

PHR	HGB - S Beta Plus Thalassemia	HGB - S Beta Zero Thalassemia	HGB - SC Sickle C Disease	HGB - SS Sickle Cell Anemia	Total	Percent of Total
PHR 1	1	1	6	13	**21**	**1%**
PHR 2	1		2	10	**13**	**1%**
PHR 3	35	10	168	295	**508**	**35%**
PHR 4	12	1	28	45	**86**	**6%**
PHR 5	7	1	15	27	**50**	**3%**
PHR 6	57	5	168	350	**580**	**40%**
PHR 7	18	4	24	57	**103**	**7%**
PHR 8	2	4	17	33	**56**	**4%**
PHR 9			4	13	**17**	**1%**
PHR 10				2	**2**	**0%**
PHR 11		1	2	6	**9**	**1%**
Total	**133**	**27**	**434**	**851**	**1,445**	100%

Source: Department of State Health Services Laboratory Information Management System

Table 4. Texas Public Health Regions (PHR) Area Map



Appendix C. Sickle Trait Cases

Table 1. Sickle Trait Cases 2008-2016 by Race/Ethnicity and Calendar Year

Year	AFRICAN AMERICAN		AMERICAN INDIAN		ASIAN		HISPANIC		OTHER		WHITE		BLANK		Total
	#	%	#	%	#	%	#	%	#	%	#	%	#	%	
2008	3,554	64	6	0.1	16	0.3	1,232	22	324	6	288	5	117	2	5,537
2009	3,485	63	4	0.1	14	0.3	1,247	23	318	6	321	6	123	2	5,512
2010	3,312	64	9	0.2	21	0.4	1,087	21	337	7	297	6	104	2	5,167
2011	3,234	64	5	0.1	17	0.3	1,048	21	337	7	301	6	127	3	5,069
2012	3,369	65	9	0.2	25	0.5	1,048	20	358	7	309	6	100	2	5,218
2013	3,525	65	8	0.1	27	0.5	1,083	20	372	7	293	5	128	2	5,436
2014	3,701	63	9	0.2	27	0.5	1,167	20	488	8	333	6	131	2	5,856
2015	3,970	65	14	0.2	34	0.6	1,130	19	491	8	323	5	109	2	6,071
2016	3,965	64	4	0.1	33	0.5	1,167	19	585	9	356	6	117	2	6,227
Total	**32,115**	**64**	**68**	**0.1**	**214**	**0**	**10,209**	**20**	**3,610**	**7**	**2,821**	**6**	**1,056**	**2**	**50,093**

Source: Department of State Health Services Laboratory Information Management System

Table 2. Sickle Trait Cases 2008-2016 by Public Health Region (PHR) and Calendar Year

PHR	2008	2009	2010	2011	2012	2013	2014	2015	2016	TOTAL
PHR 1	91	103	85	95	98	92	99	93	103	859
PHR 2	69	66	62	63	61	54	63	67	50	555
PHR 3	1,693	1,683	1,598	1,583	1,659	1,739	1,808	1,857	1,873	15,493
PHR 4	338	305	332	294	299	334	287	289	283	2,761
PHR 5	208	204	179	178	193	166	185	189	186	1,688
PHR 6	1,967	2,000	1,855	1,799	1,883	2,035	2,276	2,406	2,500	18,721
PHR 7	483	455	441	420	408	398	424	451	451	3,931
PHR 8	309	327	292	284	277	284	321	330	380	2,804
PHR 9	61	42	57	69	64	60	101	103	100	657
PHR 10	62	76	64	55	63	69	56	73	85	603
PHR 11	254	251	202	229	213	205	236	213	214	2,017
Unknown	2								2	4
TOTAL	**5,537**	**5,512**	**5,167**	**5,069**	**5,218**	**5,436**	**5,856**	**6,071**	**6,227**	**50,093**

Source: Department of State Health Services Laboratory Information Management System