# Establishment of a Sickle Cell Registry in Louisiana: Preliminary Research, Findings and Recommendations – February 2023

*There is no national surveillance of [Sickle Cell Disease (SCD)]. Consequently, there are large gaps in our current understanding of the natural course of the disease and its variable manifestations from one patient to the next, including how vulnerable populations seek and receive SCD care. The intent of the [national Sickle Cell Data Collection (SCDC)] program is to inform decisions and policies that may lead to significant improvements in the SCD community, a community that continues to be impacted by racism, bias, and prejudice.*

- United States Department of Health and Human Services, Center for Disease Control and Prevention, 2023[1]

## Introduction

Nationally and in Louisiana, it is unknown how many people are living with sickle cell disease (SCD). While SCD is the most common inherited blood disorder in the United States, there is currently no national system to collect and analyze the information needed to "drive" change in healthcare, treatment and policy to improve the health and wellbeing of individuals living with SCD.[2] Further, sickle cell trait (SCT) is more prevalent than SCD and those with SCT are also at risk for clinical complications.[3] The prevalence of SCT is also unknown. In 2022, the Louisiana legislature passed an important bill that will establish a public health registry in the state (Act 647 of the 2022 Regular Session of the Louisiana Legislature). This law is a significant step forward that will advance the Louisiana Sickle Cell Commission's long-standing call-to-action for the Louisiana Department of Health (LDH) to develop a public health monitoring system to track the number of people with SCD, their access to health care services, and health outcomes.

The purpose of this brief report is to summarize the initial research findings and recommendations related to establishing the sickle cell registry in Louisiana. As outlined in *Initiative 5: Improve Systems to Support People Living with Sickle Cell Disease* of the LDH Business Plan for State Fiscal Year (SFY) 2023, the LDH Office of Public Health (OPH) Bureau of Family Health (BFH) is leading the development of the registry. This report is the first of two that will summarize recommendations related to defining the purpose, use, operations, and technical requirements of the state's sickle cell registry.

## The Charge and Approach

Act 647 of the 2022 Regular Session of the Louisiana Legislature directs the LDH to "…establish and maintain…a registry of individuals diagnosed with sickle cell disease [in the state]" to be known as the "Skylar-Cooper Database." The purpose of this registry is to "…function as a single repository of accurate, complete records to aid in the cure and treatment of sickle cell disease…" The law outlines some foundational data reporting, data access, and rigorous protections for data collected through the registry and special studies that are also authorized by the statute. Further, the law acknowledges barriers with continuity of healthcare access that individuals with SCD experience and provides for the potential use of registry data to improve coordination and continuity of care. Lastly, the law directs LDH

[1] Department of Health and Human Services. Opportunity Number CDC-RFA-DD-23-0002: Sickle Cell Data Collection Program. Grants.gov. February 9, 2023. Accessed February 27, 2023. https://www.grants.gov/web/grants/search-grants.html?keywords=sickle%2520cell
[2] Centers for Disease Control and Prevention. Sickle Cell Data Collection Program Report: Data to Action. September 2018. Accessed February 27, 2023. https://www.cdc.gov/ncbddd/hemoglobinopathies/data-reports/2018-summer/documents/sickle-cell-data-to-action-h.pdf
[3] National Academies of Sciences, Engineering, and Medicine. 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: The National Academies Press. https://doi.org/10.17226/25632.

to establish administrative rules that will provide more specific details related to reporting, registry operations, special studies, protections and allowable uses of the data.

The approach to prepare for a sickle cell registry in Louisiana has been informed by the elements outlined in the *Guiding Framework for Setting Up a Sickle Cell Disease Surveillance System*, published by the Centers for Disease Control and Prevention (CDC). This guide was developed by CDC in partnership with states and jurisdictions that have participated in the national Sickle Cell Data Collection System (SCDC) Program and related projects that have successfully collected population-based data and have brought "data to action" to change systems of care and support for people with SCD.[4] The first steps in this framework include: establishing a multidisciplinary guidance team to guide the development of the registry; engaging members of the public with an interest in the registry to ensure that the information collected and produced by the system will be useful; and, beginning to prepare for the state rules that will define the parameters for reporting and appropriate data use.

## Preliminary Actions Taken

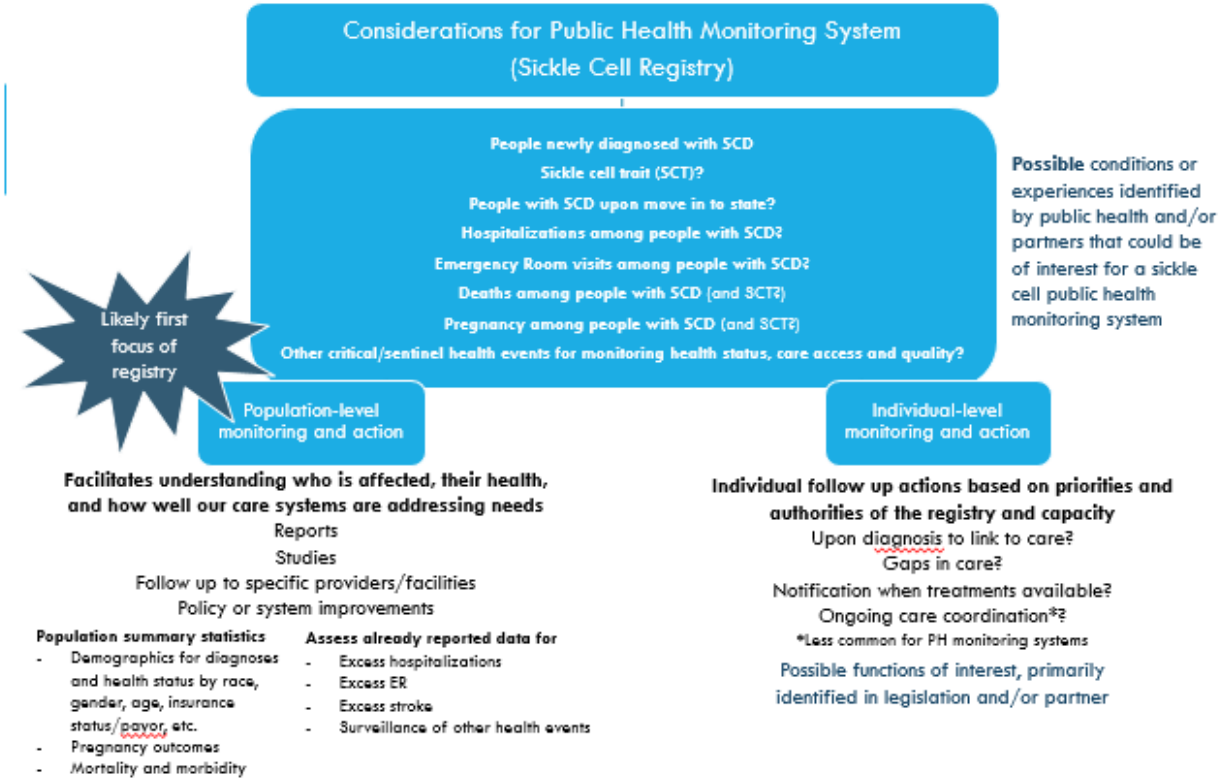
In accordance with the recommended national framework, the first work of developing the registry has been to engage others in- and outside of LDH to raise awareness of the new law and to seek input on the preliminary plans. A core internal team including Bureau leadership, the leadership over OPH BFH genetic diseases programming, and a nurse consultant has led the Bureau's work to date. Two guidance groups were convened in the fall and winter: 1) an LDH public health sickle cell guidance team, consisting of the State Health Officer, the State Epidemiologist, and the Director of the OPH Bureau of Health Informatics; and, 2) a multidisciplinary SCD Registry Steering Committee comprised primarily of external advisors including individuals with lived experience, a clinical expert, representation from one of the state's Sickle Cell Foundations (community based organization providing supportive and navigation services), a staff member from LDH Medicaid, and internal program personnel over newborn screening and sickle cell services. Lastly, OPH BFH personnel participated in the quarterly meetings of the Louisiana Sickle Cell Commission and sought input from Commission members through formal presentations at these public meetings.

Across all of these engagement activities, the focus has been to:

- Familiarize all staff and partners with the statute to establish the registry, as well as orient all to a changing context for SCD, including legislative actions taken in Louisiana over the past two years and the National Academies' *Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action* report published in 2020
- Begin to familiarize staff and partners with an understanding of public health monitoring systems (sometimes called registries, sometimes called "surveillance" systems)
- Begin to distinguish between registries that individuals voluntarily "sign up" for vs. public health registries that collect information on all individuals with certain conditions to "drive" change in policy and care systems in order to benefit all affected people and their families
- Begin to distinguish between population-level actions that public health monitoring systems can support vs. individual-level follow up and outreach activities
- Begin to explore if SCT should be monitored in addition to SCD; begin to explore types of health "events" or complications that should potentially be tracked in the registry (e.g. stroke or death)
- Identify existing data in the state related to SCD and SCT, and their allowable uses

[4] Centers for Disease Control and Prevention. Sickle Cell Data Collection Program Report: Data to Action - The SCDC Program. February 2, 2021. Accessed February 28, 2023. https://www.cdc.gov/ncbddd/hemoglobinopathies/data-reports/2018-summer/scdc-program.html

From these discussions, a preliminary conceptual framework for the registry was developed to illustrate what might be useful to include in the registry or be a product of the monitoring system. **Please note: the information in the illustration below reflects a synthesis of ideas discussed.** See the *Discussion and Preliminary Recommendations* section for considerations and anticipated next steps.




## Discussion and Preliminary Recommendations

Overall, there is consensus among the public health community and partners that there is a need for data that are complete and reliable that can characterize the health of people with SCD, and possibly SCT, in the state. There is also consensus that it will be important for data to be sufficiently rigorous to identify the strengths and gaps in the health care system to meet the needs of people with SCD, and possibly for people with SCT. However, more work is required to define the specific intended outcomes and benefits of the registry. Below are considerations for the next phases of developing the registry:

1. **Since the registry will encompass personal and sensitive information—in particular related to a condition that largely affects a population that has experienced mistreatment in the medical system—it will be critical for the registry have a clear purpose, defined allowable uses, and safeguards. This should be done before data are collected or adopted for use in the registry.**

   In general, health information is protected from intentional and unintentional disclosures by various state and federal laws, most notably the Health Insurance Portability and Accountability Act (HIPAA). In addition, the registry's authorizing legislation has specific confidentiality requirements and limitations on data uses and disclosures, including protections against legal and public records discoverability (see §1125.14 "Use of registry data" section of the law). However, any collection of personal and protected information should be guided by and limited to a well-defined purpose. The

authorizing statute states a purpose of the registry is "….to function as a single repository of accurate, complete records to aid in the cure and treatment of sickle cell disease in this state…" However, that purpose remains broad and grants the department the discretion to request "…all data and other information associated with individuals diagnosed with sickle cell disease that the secretary of the department deems necessary and appropriate for inclusion in the registry."

Anticipated actions needed:

- Review the differences between registries and surveillance systems as defined in the National Academies' *Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action*. Louisiana's "registry" may be more similar to a "surveillance system" and it will be important for terms to be well defined and clear in materials, public engagement efforts, and rules.
- Develop a logic model, possibly similar to the national Maternal Mortality Review Committee Logic Model or other format, to illustrate the purpose, uses and outcomes expected from the state Sickle Cell Disease Registry to facilitate understanding and decision-making.
- Consult CDC Sickle Cell Data Collection (SCDC) Program leadership for assistance with developing and documenting the registry's purpose, defining allowable uses, and safeguards.
- Research the population- and individual-level outcomes and benefits of registries in other states, with the assistance of the CDC SCDC Program, as needed.
- Seek legal and ethics counsel on potential proposed purpose, allowable uses, and safeguards.
- Continue to engage the LDH internal guidance team, the SCD Registry Steering Committee, the Louisiana Sickle Cell Commission, the Sickle Cell Foundations and affected individuals on the proposed purpose, allowable uses, and safeguards.
- Engage the Louisiana Hospital Association, the state's clinical systems and applicable professional associations and clinicians on the proposed purpose, allowable uses, and safeguards.

**2. As a part of developing a more specific purpose of the registry, additional discussion is required to explore what the registry should be able to do to help specific individuals who are diagnosed with SCD and SCT.**

In general, public health monitoring systems (public health "surveillance" systems) collect health-related data in order to understand how many people are living with a condition, health outcomes, deaths among individuals with the condition, trends and variations in access to care and sometimes cost of care.[5] As shown in the illustration in the previous section, this type of information can be used to inform policy changes and improvements in care systems. In the course of the discussions related to Louisiana's registry, there was also significant interest in how the data can be used to help individual people. For example, there was a need recognized for a registry to potentially identify and support outreach to people who are not accessing routine care and who might be at higher risk for life-threatening complications. Another significant example was related to ensuring that individuals with SCT are supported for their own health and in decision-making related to having children. Lastly, there was discussion of how data for public health monitoring systems usually come from clinical records and clinical providers, not from the affected individuals. Further clarification is needed around these issues.

[5] National Academies of Sciences, Engineering, and Medicine. 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: The National Academies Press. https://doi.org/10.17226/25632.

- If the registry is not voluntary or "opt-in" (most public health registries are not), it will be important to consider how the information will or could be used to help specific individuals or result in a benefit that feels significant for individuals living with SCD and SCT.
- Further discussion is needed to assess whether or not people living with SCT should also be part of the registry.

3. **Analysis of currently available data is needed in order to inform whether additional reporting will be necessary. Analysis of currently available data can also help inform and clarify the potential purposes, allowable uses, and safeguards of the registry.**

   The health department receives data from various sources that, when combined and de-duplicated, may be nearly sufficient to produce population estimates, track health outcomes and serve as data for the intended "single repository." Data currently available to the state include newborn screening test results, which include individuals with SCD and sickle cell trait, Medicaid claims data for individuals with SCD who are insured through Medicaid and who access medical care, inpatient hospital discharge data for all hospital admissions (regardless of payer), and emergency room data (regardless of payer). These are all common sources of information for states implementing SCD registries. In addition, the health department has access to and the authority to analyze birth and death records. Before potentially establishing requirements for physicians or other clinical providers to report additional information to the health department, it will be useful for currently available data to be assessed against the potential purposes, allowable uses, and safeguards identified for the registry. This one-time "data feasibility assessment" would identify alignment of current data with the goals of the registry and any gaps that may need additional data collection methodology introduced for registry completeness.

   Anticipated actions needed:
   - Secure advanced analytic capacity for data linkage and complex data analyses to assess the quality and completeness of data for supporting population estimates and ongoing public health monitoring.
   - Consult with the internal guidance team, Sickle Cell Registry Steering Committee, the Louisiana Sickle Cell Commission, the CDC, and other identified constituencies to develop the questions to guide the analytics for the one-time data feasibility assessment.
   - Document the data linkage activities, decisions and procedures used. The *Guiding Framework for Setting Up a Sickle Cell Disease Surveillance System* outlines specific activities that will be required to develop the data infrastructure. Documenting the approaches used and results will facilitate future decision-making.
   - Publish a profile SCD and SCT in the state based on available data with information about the timeliness, accuracy, completeness and feasibility of using data from the various existing data sources to achieve the required and proposed purposes of the registry.

## Next Steps

As outlined in the LDH Business Plan for State Fiscal Year (SFY) 2023, the primary focus of efforts related to the development of the registry in the coming months will be to engage with partners in and out of the state to refine the purpose and potential uses for the registry. Public comment will be open through April 15, 2023 on https://ldh.la.gov/page/SickleCellDisease_SCD_Louisiana.