

SICKLE CELL ANNUAL REPORT 2019



Ohio

Department
of Health

Ohio Sickle Cell Services Program 2019 Annual Report

This report includes data from state fiscal year 2018

SUBMITTED TO

Members of the 133rd General Assembly

SUBMITTED BY

Ohio Department of Health
Sickle Cell Services Program



Department
of Health

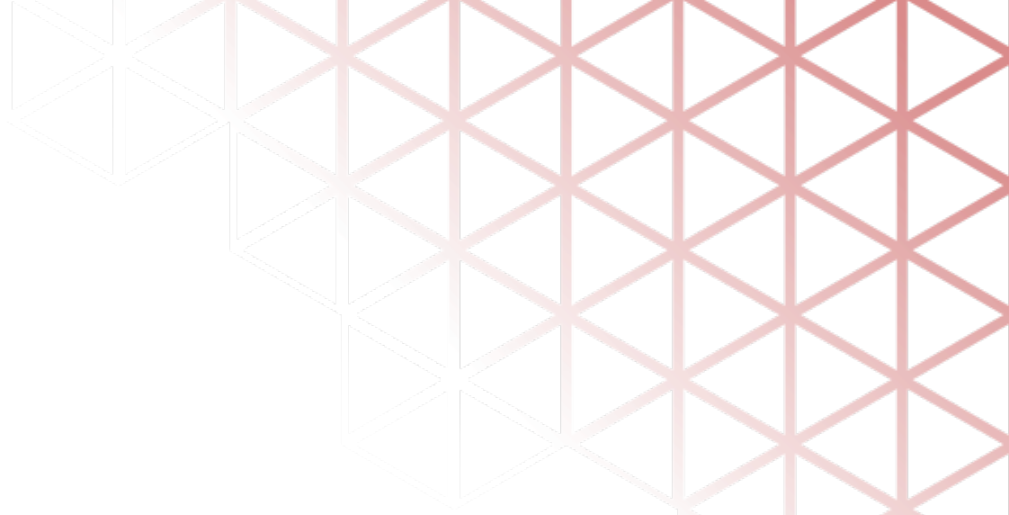


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We are especially appreciative of the data analysis provided by Hannah MacDowell (ODH Research and Evaluation) and the design services provided by Janet Steadman (ODH Public Affairs and Communications) in the development of this report.

A special acknowledgement and thank you is also extended to the project directors and staff at the Ohio state-funded sickle cell projects. This report is made possible by their dedication and “hands-on” participation in the completion of required program data collection and reporting forms during the state fiscal year (SFY).

Most importantly, we would like to thank the many patients and families at the Ohio pediatric and adult sickle cell treatment centers and the affected family support group members for their continuous contributions to our education.

From the Director

Dear Members of the 133rd General Assembly:

On behalf of the patients and families that we serve, I am pleased to submit to you the annual report for the Ohio Department of Health Sickle Cell Services Program.

Under [Section 3701.131 of the Ohio Revised Code](#), requirements are set forth for the Director of Health to:

“Encourage and assist in the development of programs of education and research pertaining to the causes, detection, and treatment of sickle cell disease and provide for rehabilitation and counseling of persons possessing the trait or afflicted with this disease...”

In accordance with the above, this annual report to the General Assembly serves to outline the receipt and disbursement of funds and the implementation and progress of various programs undertaken pursuant to this section.

Should you have any questions or need additional information, please contact Lisa Griffin, Director of Government Affairs by phone at (614) 644-9164 or email at lisa.griffin@odh.ohio.gov.

Sincerely,



Amy Acton, MD, MPH
Director
Ohio Department of Health



Executive Summary

In SFY 2018 (July 1, 2017-June 30, 2018), the Ohio Department of Health (ODH) funded a statewide sickle cell project (SWSCP) located in Columbus and six regional sickle cell projects (RSCPs) located Cincinnati, Dayton, Toledo, Columbus, Cleveland, and Akron. The projects are responsible for collecting data pertaining to the level and scope of sickle cell services in their respective regions or statewide for the reporting SFY. The data in this report is compiled from SFY 2018 Minimum Data Set collection forms and other program reporting formats.

Newborn Hemoglobinopathy Screening

- The RSCPs received notification of 3,553 abnormal newborn hemoglobinopathy screening results from the ODH Public Health Laboratory during SFY 2018. This figure accounts for 2.6% of all Ohio births (n=137,588) during this reporting period.
 - 129 (4%) newborns had a disease hemoglobinopathy screening result, and 3,424 (96%) had a carrier hemoglobinopathy screening result.

Hemoglobinopathy Testing

- The RSCPs reported confirmatory testing on 2,102 (59%) of newborns identified in SFY 2018 with an abnormal newborn hemoglobinopathy screening result. Documentation of confirmatory testing was not obtained on 1,451 (41%) newborns with an abnormal screening notification due to the parent/guardian being lost to follow-up, the physician of record not reporting to the RSCP, the infant's parent/guardian declining RSCP services or the infant expiring before testing.
- Of the 129 with a disease hemoglobinopathy screening result:
 - 109 (85%) received confirmatory testing within two months of life and 20 (15%) received confirmatory testing after two months.
 - 57 (44%) received treatment within three months of birth and one (1%) received treatment after three months. Sixty-eight (53%) newborns did not receive treatment either due to being confirmed as carriers or having diseases not requiring treatment. Three (2%) newborns had an unknown treatment status.
- Three-quarters of newborns (n=1,581) with confirmatory testing for hemoglobinopathies identified as Black or African-American.
- A total of 2,098 non-newbornsⁱ received hemoglobinopathy testing services through an RSCP in SFY 2018, with 796 (38%) identified with a carrier or disease hemoglobinopathy. A total of 1,302 (62%) non-newborns were identified with no hemoglobinopathy.
- Over two-thirds (67%, n=1,408) of non-newborns receiving hemoglobinopathy testing services identified as Black or African-American.
- Of those who received hemoglobinopathy testing services, 2,101 (50%) were newborns from 54 counties and 2,098 (50%) were non-newborns from 34 counties in Ohio.

Hemoglobinopathy Counselingⁱⁱ

- Of the 2,102 newborns born in SFY 2018 who received confirmatory testing, 1,971 (94%) had a parent or guardian receive hemoglobinopathy counseling by the RSCPs.
 - There were 3,650 parents or guardians who received hemoglobinopathy counseling in association with these newborns.
 - For the 131 confirmed newborns whose parents or guardians did not receive counseling, 89% (n=116) were due to the caregivers being lost to follow-up, 9% (n=12) were due to the caregivers declining counseling and 2% (n=3) had an unknown counseling status.
- Hemoglobinopathy counseling services were provided by the RSCP to 1,965 (94%) of the non-newborns who received diagnostic testing in SFY 2018.

Hemoglobinopathy Outreach Educationⁱⁱⁱ

- Sickle cell project staff provided 779 education events, totaling 2,181 hours of direct education to an estimated 23,181 public/community members, health care providers, and community professionals.
 - Most education events took place in health care settings (66%, n=518) and schools (18%, n=140).
 - The most common educational topics were general hemoglobin disease and trait overview (n=702), clinical/treatment/management issues (n=377), and newborn hemoglobin screening (n=357). Events often covered more than one topic.
 - Most education events were in the format of a presentation (34%, n=266), training (32%, n=253) or small group instruction (28%, n=219).
 - Education events were held in 18 Ohio counties. This included the five counties in Ohio with the highest proportion of African-American residents (Cuyahoga, Hamilton, Franklin, Montgomery, and Lucas).
- In addition, project staff conducted 222 awareness activities, with an estimated 15,155,389 contacts made raising awareness of sickle cell disease and other hemoglobinopathies.
 - The top three modes of awareness were community or professional outreach and engagement (55%, n=121), material distribution (18%, n=39), and social media (13%, n=28).
 - During Sickle Cell Sabbath, an interfaith outreach campaign designed to educate and increase awareness in the faith community, sickle cell project staff reported reaching 365 faith-based organizations in the state.

ⁱ Non-newborn is defined as an individual born before SFY 2018 (07/01/17-06/30/18) who received services through the RSCP or non-RSCP provider during the SFY 2018.

ⁱⁱ Hemoglobinopathy counseling refers to the entire process of communicating accurate test results to individuals identified with a hemoglobin variant. This counseling process involves the clear communication of the medical, psychological, social, and genetic factors related to the condition being discussed.

ⁱⁱⁱ Hemoglobinopathy Outreach Education focuses primarily on efforts to 1) increase the awareness, knowledge and skill level of Ohio professionals about the special health care needs and services related to hemoglobinopathies, and 2) expand public and community education and awareness of hemoglobinopathies.



Introduction

Program Background

The [Ohio Revised Code, under Section 3701.131](#) sets forth requirements for the Director of Health to:

“(A) Encourage and assist in the development of programs of education and research pertaining to the causes, detection, and treatment of sickle cell disease and provide for rehabilitation and counseling of persons possessing the trait of or afflicted with this disease;”

“(B) Advise, consult, cooperate with, and assist, by contract or otherwise, agencies of this state and the federal government, agencies of the governments of other states, agencies of political subdivisions of this state, and private organizations, corporations, and associations in the development and promotion of programs pertaining to the causes, detection, and treatment of sickle cell disease and rehabilitation and counseling of persons possessing the trait of or afflicted with this disease;”

“(C) Accept and administer grants from the federal government or other sources, public or private, for carrying out any of the functions enumerated in divisions (A) and (B) of this section.”

In addition, [Section 3701.501 of the Ohio Revised Code](#) requires that:

“... all newborn children shall be screened for the presence of the genetic, endocrine, and metabolic disorders specified in rules adopted pursuant to this section.”

To carry out these requirements, ODH funded two grant initiatives in SFY 2018 under the Sickle Cell Services Program related to sickle cell disease (SCD), sickle cell trait (SCT), and other hemoglobinopathies:

The **Sickle Cell Initiative** is comprised of a regional network of six sickle cell projects based in four pediatric hospitals and two community-based agencies located in Cincinnati, Dayton, Toledo, Columbus, Cleveland, and Akron (see Appendices B through G). These projects are funded to ensure the provision of comprehensive sickle cell services for newborns, children, and adults, including:

- Newborn screening coordination and follow-up of abnormal hemoglobin disease and hemoglobin trait results.
- Hemoglobinopathy counseling and education of patients and their families.
- Public and professional outreach/education/awareness activities and resource materials.
- Referral services to specialized medical teams for hemoglobin disease management.

The **Statewide Family Support Initiative** project, located in Columbus (see Appendix H), is funded to support the provision of statewide training, education, and empowerment resources to individuals (adults/young adults) and families at risk or affected by SCD, SCT, and other hemoglobinopathies and to the professionals who serve them. These services include:

- Statewide public awareness and media campaigns.
- Professional training and outreach.
- Consumer support/empowerment resources.
- Advisory organization for the Ohio Sickle Cell Affected Family Association.
- Linkage with the National Sickle Cell Disease Organization – [Sickle Cell Disease Association of America, Inc.](#) (SCDAA).

Receipt and Disbursement of Funds

In accordance with [section 3701.501 of the Ohio Revised Code](#):

“The director shall adopt rules in accordance with Chapter 119. of the Revised Code establishing a fee that shall be charged and collected in addition to or in conjunction with any laboratory fee that is charged and collected for performing the screenings required by this section. The fee, which shall be not less than fourteen dollars, shall be disbursed as follows...”

“Not less than three dollars and seventy-five cents shall be deposited into the state treasury to the credit of the sickle cell fund, which is hereby created. Money credited to the sickle cell fund shall be used to defray costs of program authorized by [section 3701.131 of the Revised Code](#).”

For SFY 2018, fees generated from the sale of newborn screening kits to Ohio hospitals and birthing facilities were utilized by ODH to award \$800,000 in grant subsidies per year (see Table 1).

- The six sickle cell projects located in Cincinnati, Dayton, Toledo, Columbus, Cleveland and Akron were awarded a total of \$710, 000 to provide services/activities (see Program Background) under the **Sickle Cell Initiative**.
- The sickle cell project located in Columbus was awarded \$90,000 to provide services/activities (see Program Background) under the **Statewide Family Support Initiative**.

Funding to the sickle cell projects for SFY 2018 under both the **Sickle Cell Initiative** and the **Statewide Family Support Initiative** were continuation grant awards distributed through an established ODH grant application and selection process.

Table 1. Grant Award Allocations

| Sickle Cell Initiative | | | |
|--|---|---------------------|-----------------------|
| Region | Agency Name | City/County | SFY 2018 Award |
| I | Cincinnati Children’s Hospital Medical Center | Cincinnati/Hamilton | \$125,957 |
| II | Children’s Medical Center-Dayton | Dayton/Montgomery | \$72,932 |
| III | Neighborhood Health Association of Toledo, Inc. | Toledo/Lucas | \$54,743 |
| IV | Nationwide Children’s Hospital | Columbus/Franklin | \$171,595 |
| V | American Sickle Cell Anemia Association | Cleveland/Cuyahoga | \$188,164 |
| VI | Children’s Hospital Medical Center-Akron | Akron/Summit | \$96,609 |
| Statewide Family Support Initiative | | | |
| | Ohio Sickle Cell and Health Association | Columbus/Statewide | \$90,000 |



Program Implementation

Newborn Hemoglobinopathy Screening

“Newborn screening programmes are comprehensive systems which provide an essential public health service. Their mission is to eliminate or reduce the mortality, morbidity and disability that result from congenital disease by ensuring that all affected infants receive early diagnosis and long-term treatment in an attempt to achieve optimal health, growth and development”.¹

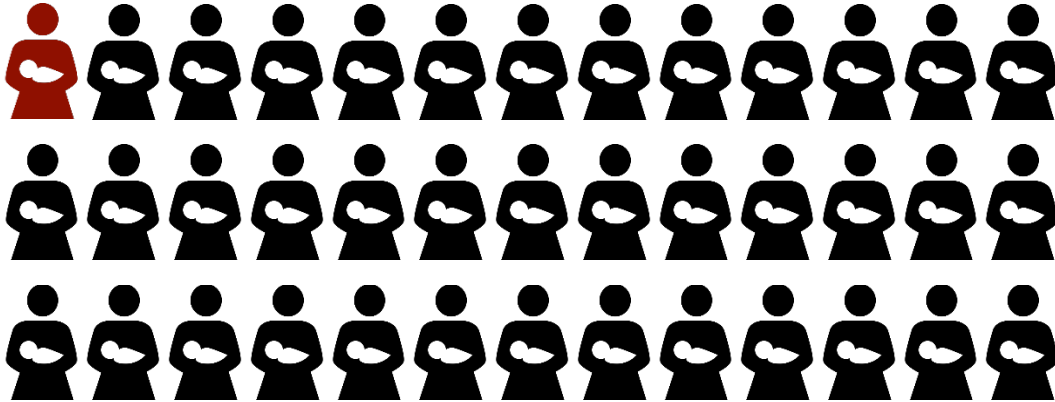
Since March 1990, all infants born in the state of Ohio are screened at birth for SCD, SCT, and other hemoglobinopathies as part of the newborn bloodspot screenings mandated by Ohio Administrative Code. The primary purpose of hemoglobinopathy screening is to identify infants with a SCD (e.g., HbSS, HbSC, HbS β^0 -thalassemia, HbS β^+ -thalassemia) and initiate penicillin prophylaxis, which has been shown to substantially reduce the incidence of pneumococcal sepsis in infancy.² Early identification, when linked to timely diagnostic testing, parental education, and comprehensive care, can markedly reduce morbidity and mortality for SCD in infancy and early childhood. At present all 50 states, District of Columbia, and the U.S. territories require and provide universal newborn screening (NBS) for SCD and other hemoglobinopathies.

Newborn hemoglobinopathy screening also identifies infants with other hemoglobinopathies, such as “carriers” of hemoglobin variants. While not clinically significant, detection of a carrier state (e.g., SCT), provides a “genetic” window into the family that can result in the identification of couples at risk for having children with SCD in subsequent pregnancies. It may also identify other family members at risk or affected by SCD, SCT, or a related hemoglobinopathy. Thus, the value of trait identification is threefold: (1) educate families; (2) test other family members, especially those who are at reproductive age and who may not have been screened at birth; and (3) provide hemoglobinopathy counseling.

All bloodspot screenings performed in Ohio are analyzed by the ODH Public Health Laboratory under the [Newborn Screening Program](#). The RSCPs, under the Sickle Cell Initiative, are an integral part of this program. They are directly responsible for case-by-case follow-up of all presumptive positive abnormal hemoglobin results (hemoglobin disease and hemoglobin trait) reported by the NBS Program to the newborns’ physician of record. Follow-up of abnormal newborn hemoglobin screening results is in accordance with [Chapter 3701-55 of the Ohio Administrative Code governing Genetic, Endocrine or Metabolic Screening of Newborn Infants](#). This follow-up includes assisting with and/or providing a point of referral for confirmation, consultation, education, counseling, and medical management of those newborns identified with an abnormal hemoglobin result.

During SFY 2018, the RSCPs received notification of 3,553 abnormal newborn hemoglobinopathy screening results (from the ODH Public Health Laboratory). This figure accounts for 2.6% of all Ohio births (n=137,588) during this reporting period (see Figure 1).

Figure 1. Proportion of Newborns with an Abnormal Hemoglobinopathy Screening Notification



Sources: Ohio Public Health Information Warehouse, Birth Occurrence (SFY 2018 births); Minimum Dataset SFY 2018 Table 1

Hemoglobinopathy Testing

Confirmation of a presumptive positive hemoglobinopathy screening result is an integral part of the NBS follow-up process. In addition to confirmation of newborns with an abnormal hemoglobin result, the RSCPs may also provide diagnostic testing services to individuals at risk for hemoglobinopathies. These individuals, referred to as “non-newborns,” include:

- Parents and family members of infants identified with abnormal NBS results.
- Adults (not associated with an abnormal NBS result) unaware of their hemoglobin status.
- Individuals of childbearing age (representing various racial and ethnic populations) considered to be at increased risk for hemoglobinopathies.
- Children whose parents do not know if they are trait positive.
- Student-athletes going to college (requirement of the National Collegiate Athletic Association - NCAA).

Newborn Hemoglobinopathy Testing

The RSCPs received notification of 3,553 abnormal hemoglobinopathy screening results for newborns born in SFY 2018. Of these newborns, the RSCPs reported 2,102 (59%) received confirmatory testing (see Figure 2). The reasons newborns were not documented as having confirmatory testing is described in Table 2.

Figure 2. Newborns – Abnormal Screening Notification and Confirmatory Testing

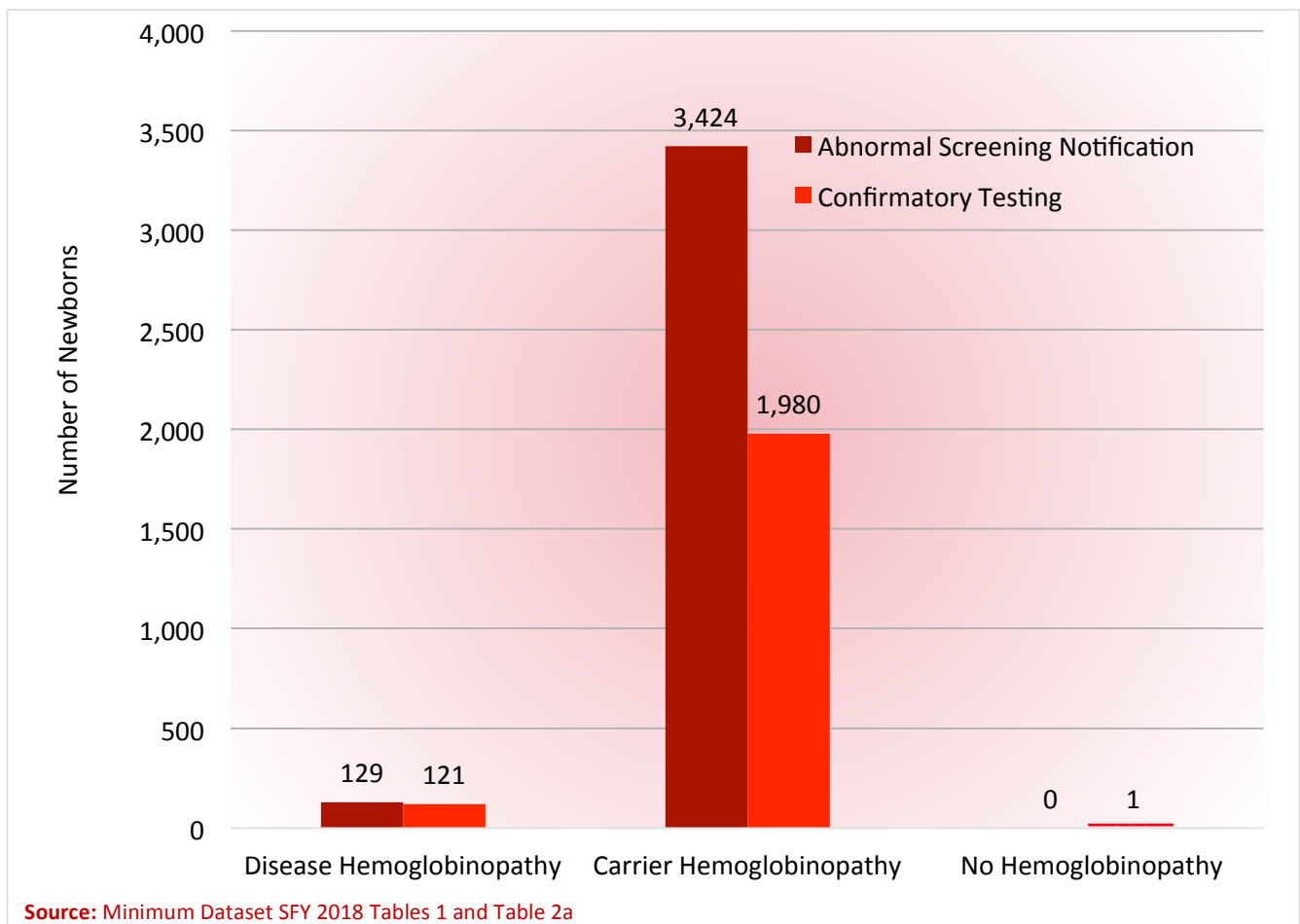


Table 2. Reasons Newborns were not Documented as having Confirmatory Testing

| Reason | No. of Newborns (n=1,451) | Percent of Newborns |
|--|--------------------------------------|----------------------------|
| Parent/guardian being lost to follow-up | 818 | 56.4% |
| Physician of record did not report to RSCP | 606 | 41.8% |
| Parent/guardian declined services | 21 | 1.4% |
| Newborn expired | 6 | 0.4% |

Source: Minimum Dataset Table 2b

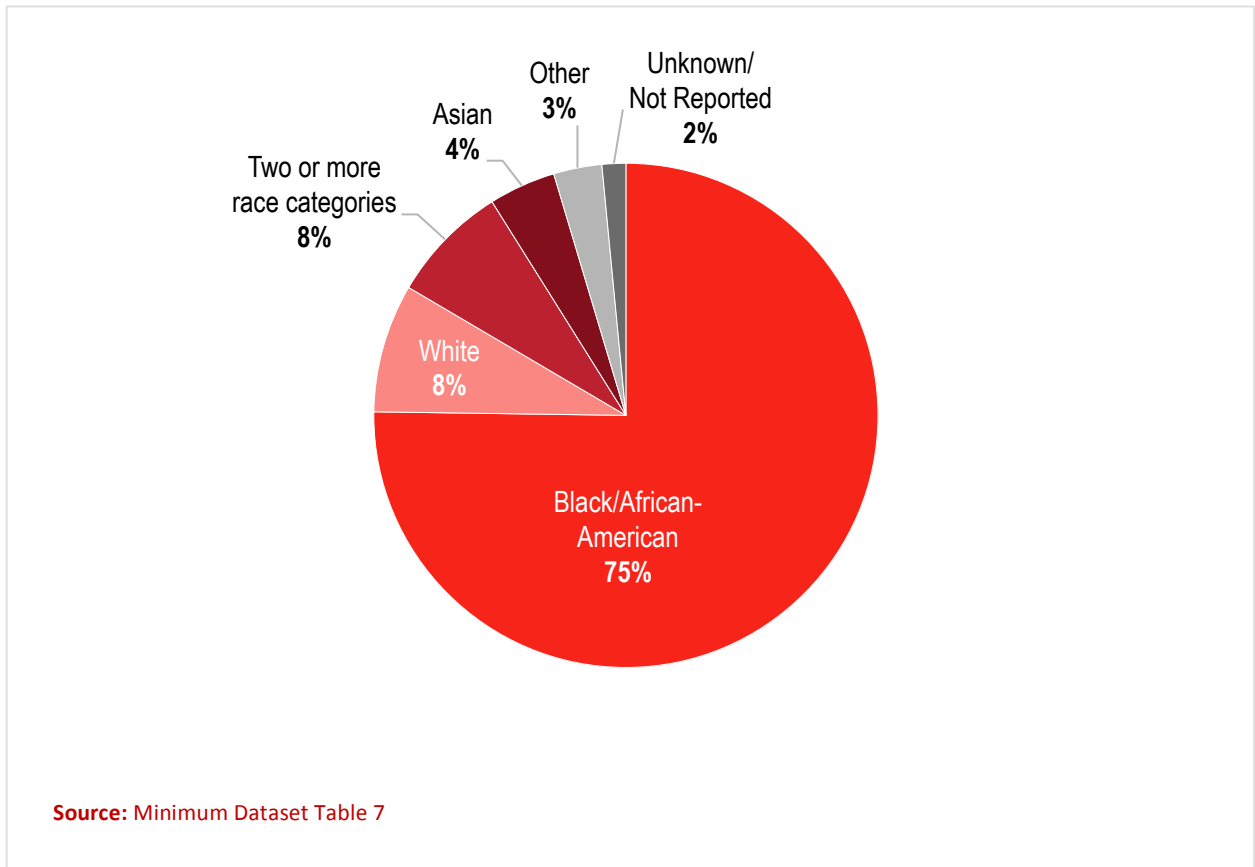
Time from Birth to Diagnosis and Treatment

During SFY 2018, the RSCPs received notification of 129 abnormal newborn screening results indicating a disease hemoglobinopathy. Of these, 109 (85%) received confirmatory testing within the first two months of life and 20 (15%) received confirmatory testing after two months.

Newborns with treatable hemoglobinopathies should begin treatment (initiation of penicillin prophylaxis for SCD is a standard of care) within the first three months of life. Of the 129 newborns with a disease hemoglobinopathy screening result, RSCPs reported 57 (44%) newborns receiving treatment within three months of birth and one (1%) receiving treatment after three months. Sixty-eight (53%) newborns did not receive treatment either due to being confirmed as carriers or having diseases not requiring treatment. Three newborns (2%) had an unknown treatment status; two were due to noncompliance of the caregivers, and one was due to the family moving outside of the country.

Three-quarters (n=1,581) of newborns with confirmatory testing identified as Black or African-American (see Figure 3). The category of “Other” in Figure 3 includes the race classifications of Pacific Islander, American Indian/Alaskan Native, and Other.

Figure 3. Newborns with Documented Confirmatory Testing - Race (n=2,102)

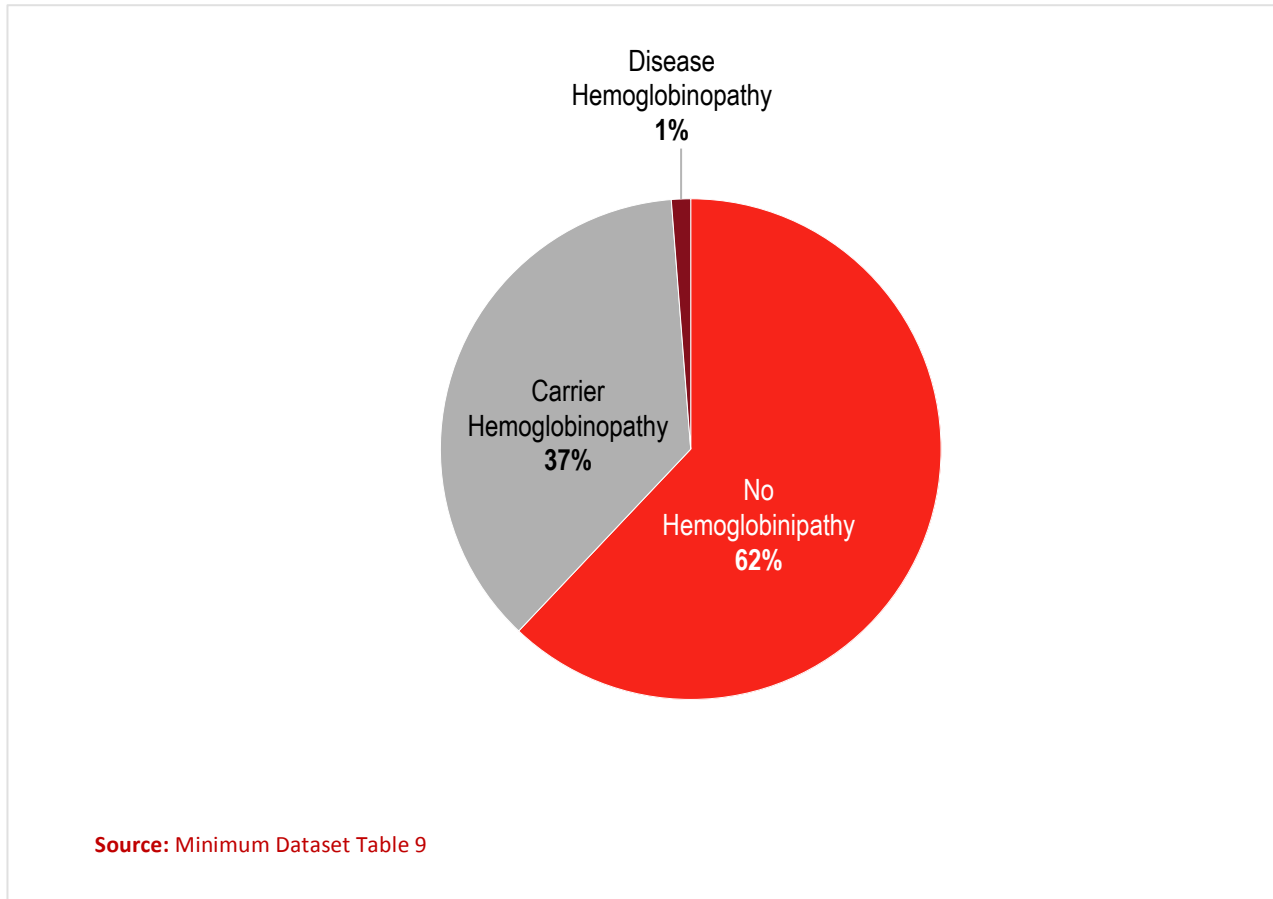


Non-Newborn Hemoglobinopathy Testing

In SFY 2018, 2,098 non-newborns received hemoglobinopathy testing services through an RSCP, with 26 (1%) identified with a disease hemoglobinopathy and 770 (37%) identified with a carrier hemoglobinopathy. Slightly more than half (n=1,054) of the non-newborns who received testing were 21 years old or younger and less than half (n=1,044) were over 21 years of age.

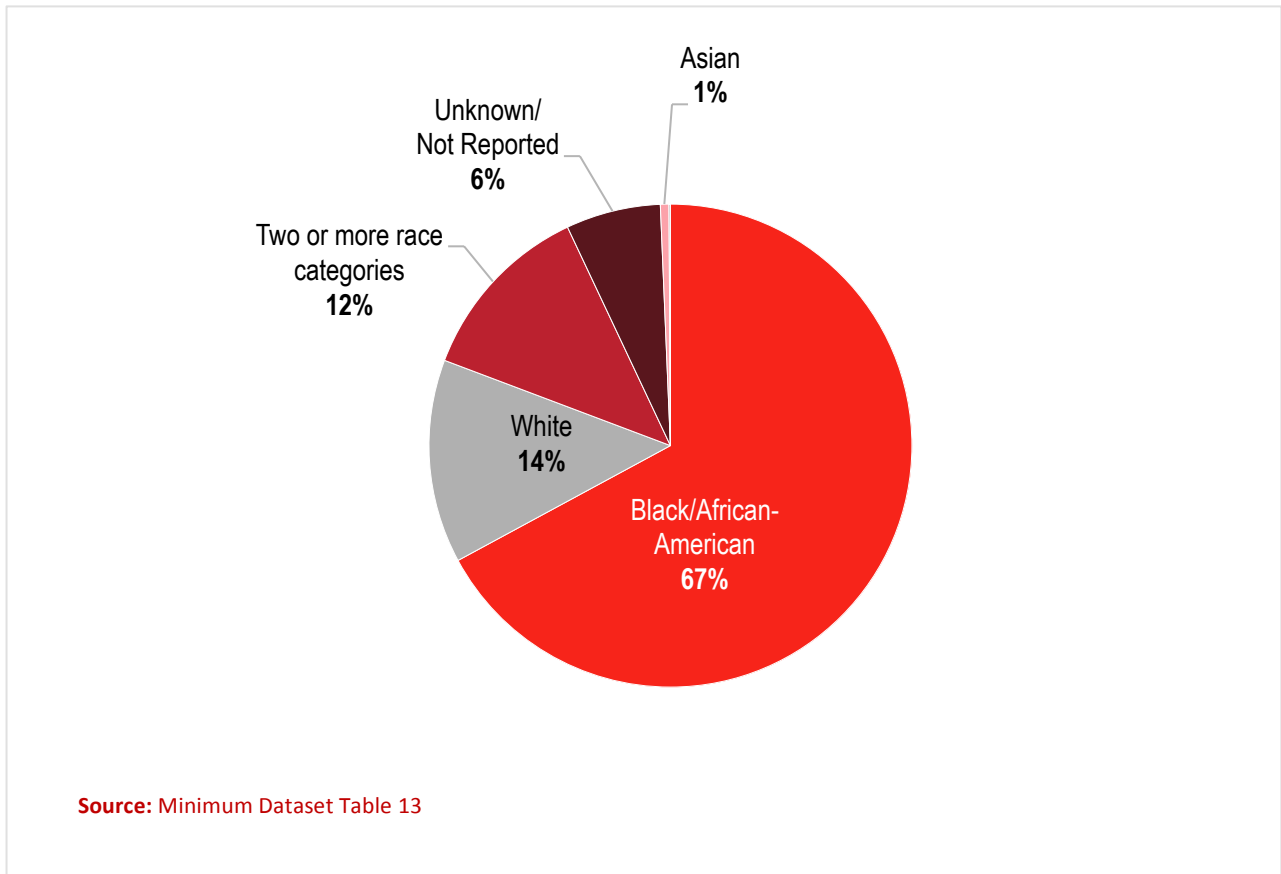
A total of 1,302 (62%) non-newborns were identified with no hemoglobinopathy.

Figure 4. Non-Newborns with Hemoglobinopathy Testing (n=2,098)



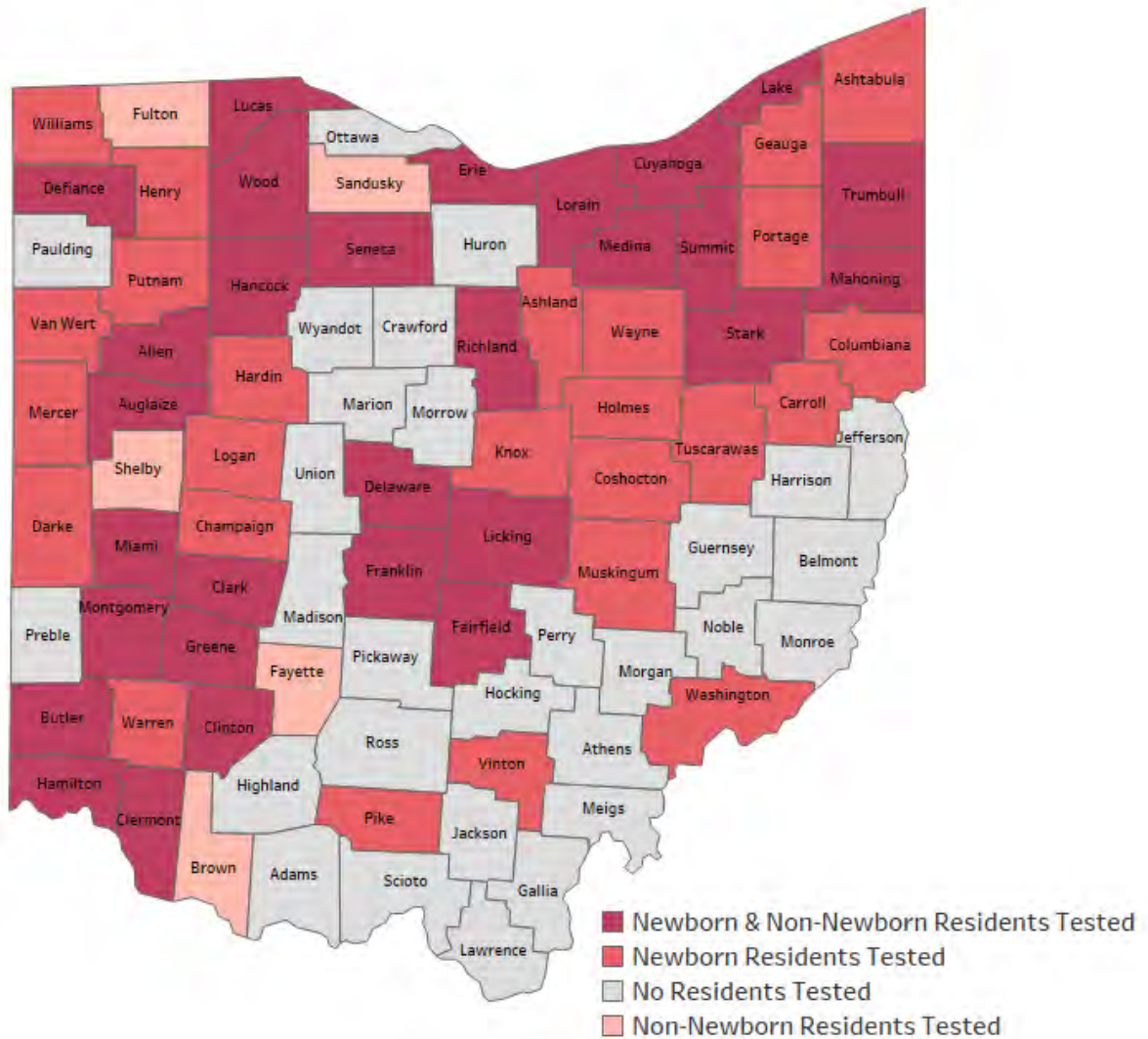
More than two-thirds (n=1,408) of non-newborns receiving hemoglobinopathy testing services identified as Black or African-American (see Figure 5).

Figure 5. Non-Newborns with Hemoglobinopathy Testing - Race (n=2,098)



Of those who received hemoglobinopathy testing services, 2,101 (50%) were newborns from 54 counties and 2,098 (50%) were non-newborns from 34 counties in Ohio (see Figure 6). One newborn who received testing resided out of state.

Figure 6. Newborns and Non-Newborns with Hemoglobinopathy Testing - County



Source: Minimum Dataset Table 8 and Table 14

Hemoglobinopathy Counseling

Counseling related to the diagnosis of a hemoglobinopathy is a vital link to the successful utilization of comprehensive, specialized clinical health care services provided through the Ohio network of pediatric and adult treatment centers.

Hemoglobinopathy counseling services provided by the RSCPs are offered on a regional basis to the parents and/or family members of all infants identified and confirmed with an abnormal newborn hemoglobinopathy screening result and “non-newborns” identified through various referral sources including physician and other health care providers, community providers, ODH partner programs, and those that are self-identified.

Children with a diagnosis of SCD, SCT, or other hemoglobinopathy, confirmed outside of the newborn period, are also offered hemoglobinopathy counseling services upon referral and/or request.

In addition, non-RSCP providers (providers external to the RSCP) may choose to counsel parents of newborns with a confirmed abnormal hemoglobin result at their primary location and not refer to the RSCP.

Newborns

Of the 2,102 newborns born in SFY 2018 who received confirmatory testing, 1,971 (94%) had a parent or guardian receive hemoglobinopathy counseling. In association with these newborns, RSCPs documented 3,650 parents or guardians who received hemoglobinopathy counseling. For 131 confirmed newborns (six confirmed with disease, 125 confirmed with trait) whose parents or guardians did not receive counseling, 89% (n=116) were due to the caregivers being lost to follow-up, 9% (n=12) were due to the caregivers declining counseling, and 2% (n=3) were unknown.

Non-Newborns

Hemoglobinopathy counseling services were provided in association with 1,965 (94%) non-newborns who received diagnostic testing.

Hemoglobinopathy Outreach Education

Although sickle cell disease is a common genetic disorder, there still exists today a considerable lack of knowledge and awareness of the disorder.

As one of its primary goals, the ODH Sickle Cell Services Program, in partnership with the network of state-funded sickle cell projects, promotes and disseminates information about hemoglobinopathies and related services to health care and community professionals/providers, the general public, and at-risk^{iv} segments of the community.

The implementation and delivery of education services at the project level is of primary importance because of its direct impact on the quality and accessibility of health care services for the individuals at risk or affected by hemoglobinopathies, and the competency of practitioners involved in these services. Educational services are an integral part of each project's objective to reach these targeted audiences.

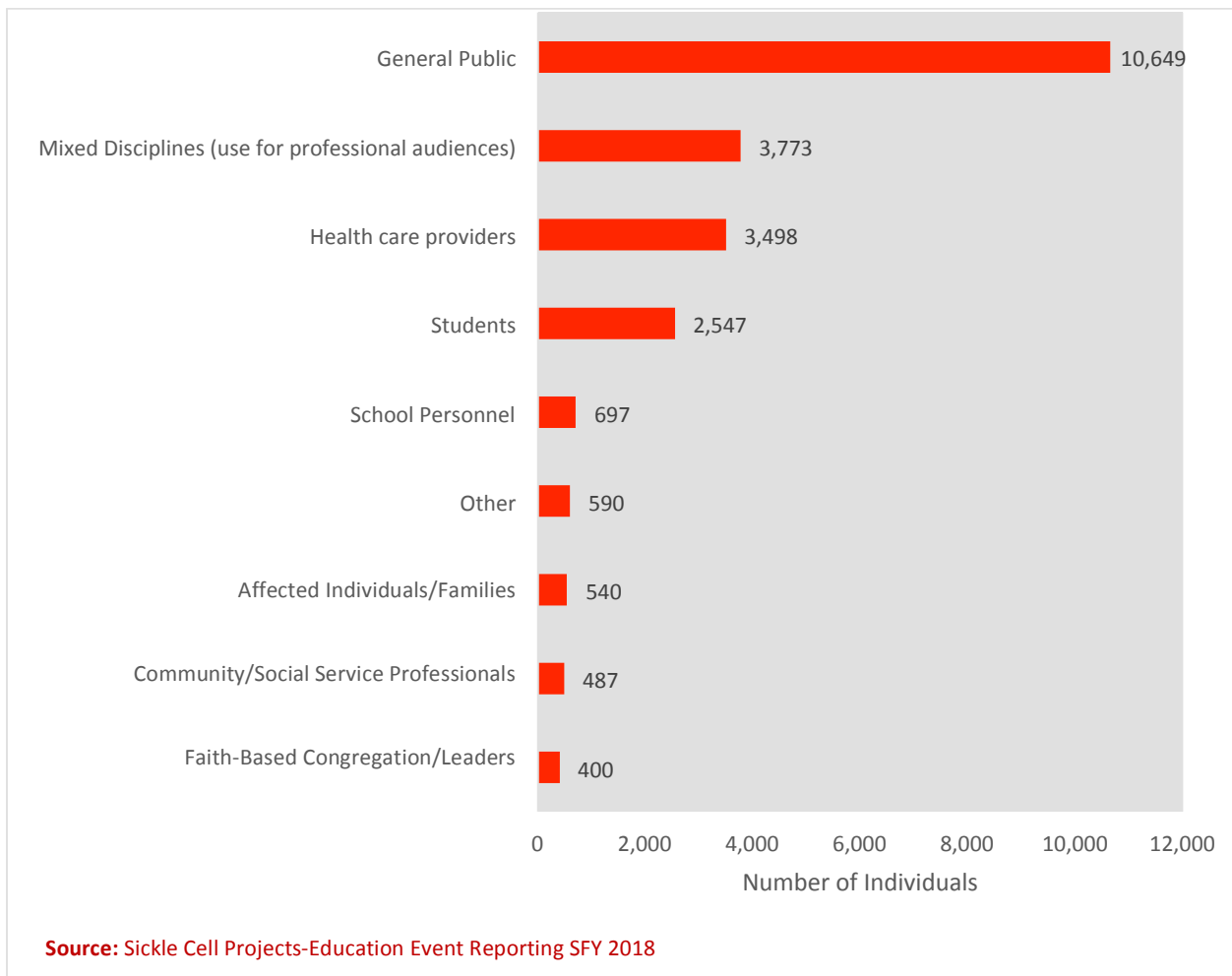
Sickle cell awareness campaigns (e.g., September-National Sickle Cell Awareness Month and Sickle Cell Sabbath) are another aspect of a larger program of professional, patient, and public education designed to reduce the risk and consequence of SCD and other related hemoglobinopathies. During the SFY, the sickle cell projects carry out awareness activities to inform the public about the disease and increase visibility of sickle cell related services and resources.

Throughout the year, the projects also engage SCD-interested organizations and stakeholders to further promote awareness of SCD and educate the public by disseminating current, up-to-date information and key messages to increase healthy outcomes among various racial, ethnic, and age groups related to hemoglobinopathies.

^{iv} At-risk segments of the community include individuals with heritage from sub-Saharan Africa; Spanish-speaking regions in the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India, and Mediterranean countries such as Turkey, Greece and Italy.

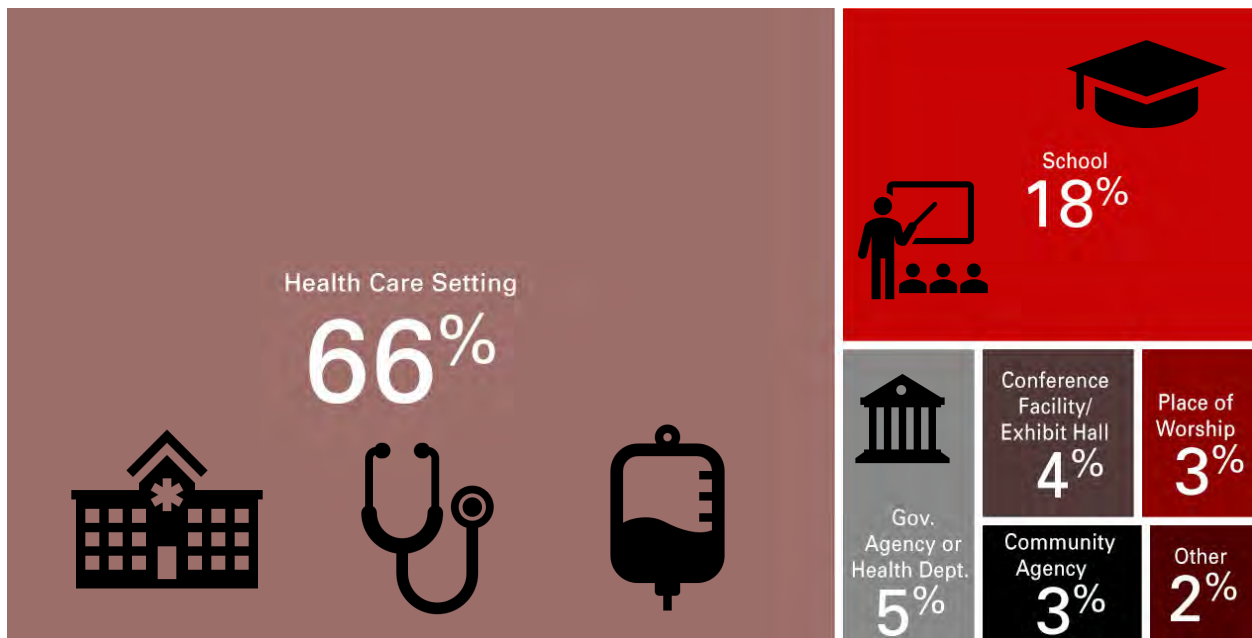
During SFY 2018, funded projects conducted a total of 779 education events, which provided education to an estimated 23,181 individuals (see Figure 7). The majority of events were provided to the general public, professionals from mixed disciplines, health care providers, and school student audiences, respectively. In total, the projects reported over 2,181 hours of direct education.

Figure 7. Education Events by Audience Type



Education events took place in a variety of settings, with most taking place in health care settings (66%, n=518) and schools (18%, n=140). In Figure 8, health care settings included pediatric and adult hospitals, medical provider offices, and health centers. School settings included daycare, preschool, pre-kindergarten, grades K-12, and vocational/post-secondary institutions. The category of “Other” included business, outdoor facility or recreation centers, and other settings (as specified by the RSCPs and SWSCP).

Figure 8. Education Events by Facility Type (n=779)



Source: Sickle Cell Projects-Education Event Reporting SFY 2018

The most common educational topic was general hemoglobin disease and trait overview (n=702), followed by clinical/treatment/management issues (n=377) and newborn hemoglobin screening (n=357). Events often covered more than one of the topics in Figure 9.

Figure 9. Education Events by Topic

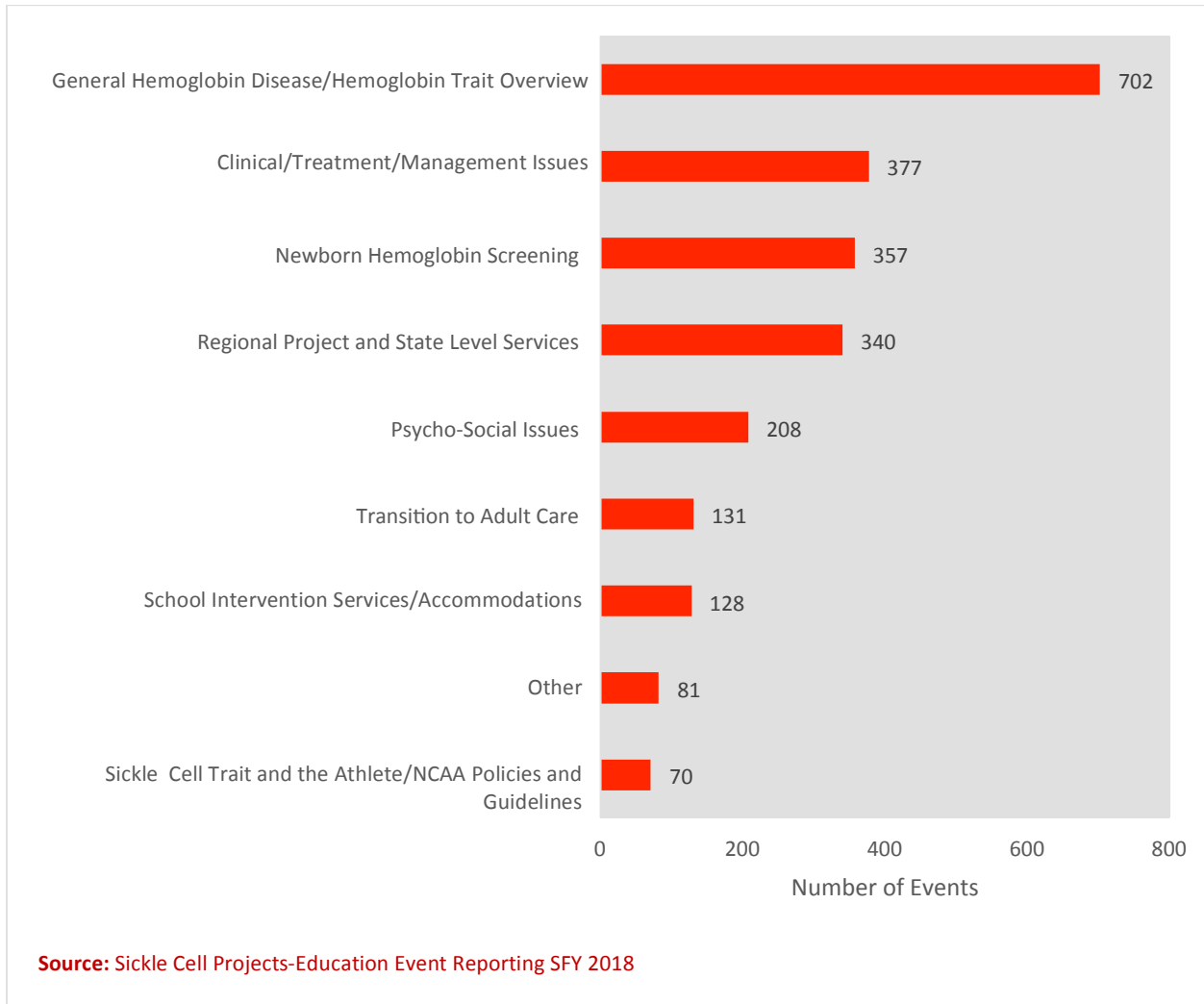
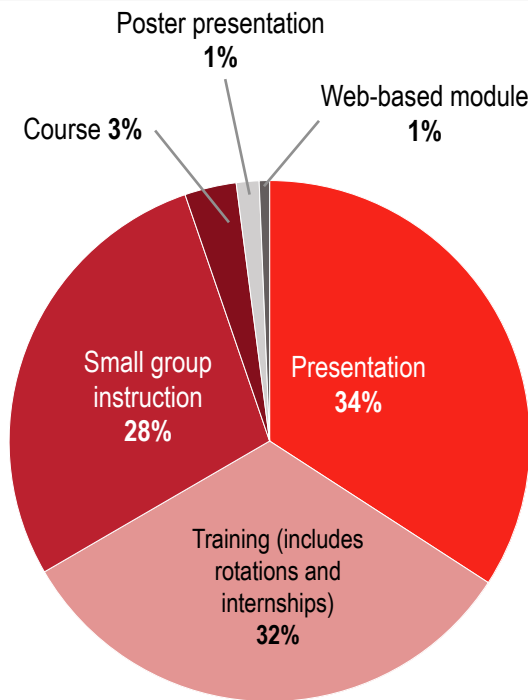


Figure 10 shows that most education events were in the format of a presentation (34%, n=266), followed by training (32%, n=253) and small group instruction (28%, n=219).

Figure 10. Education Events by Instructional Format (n=779)

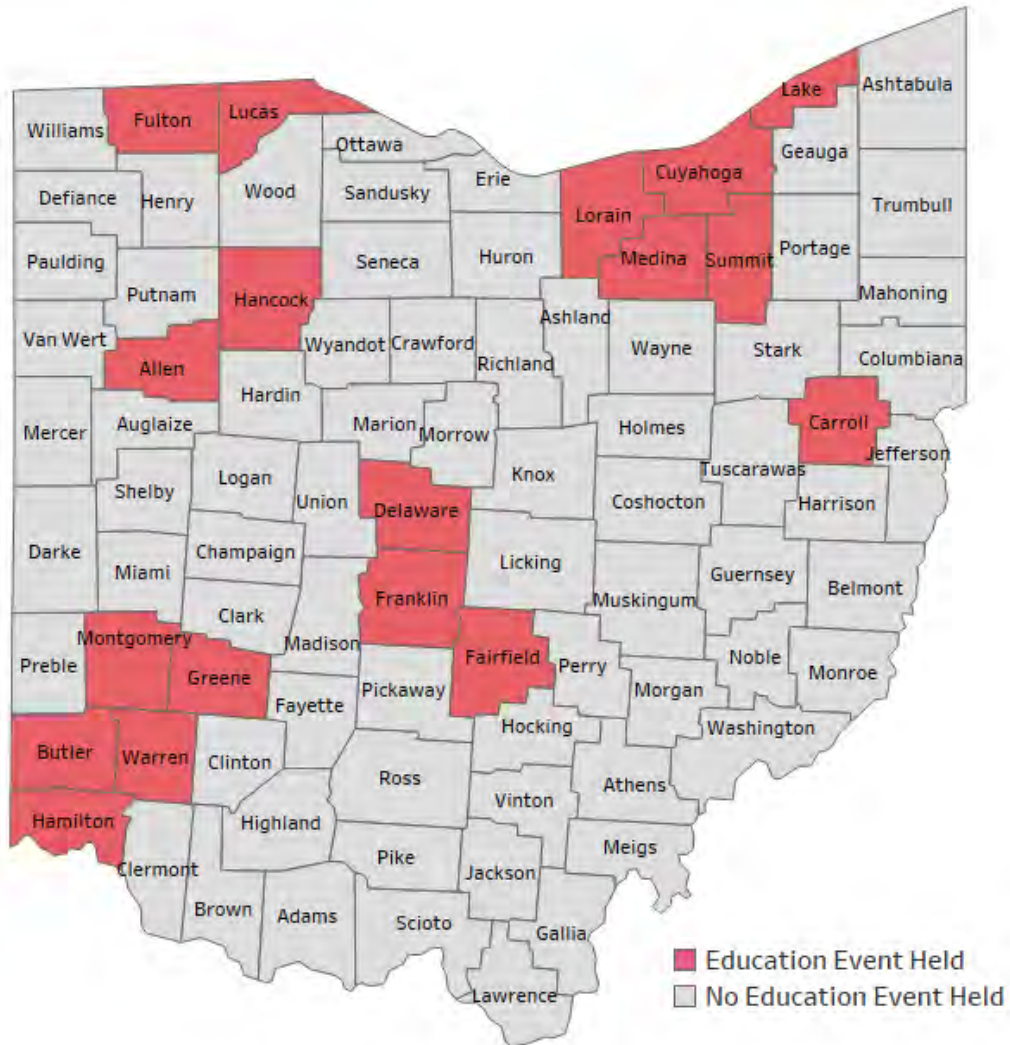


Source: Sickle Cell Projects-Education Event Reporting SFY 2018

Education events took place in 18 Ohio counties (see Figure 11). This included the five counties with the highest proportion of African-American residents (Cuyahoga, Franklin, Hamilton, Montgomery, and Lucas).³

Five events were web-based modules and could have reached audiences beyond these counties.^v

Figure 11. Education Events by County



Source: Sickle Cell Projects-Education Event Reporting SFY 2018

^v For SFY 2018, data on awareness/media activities in Ohio was not collected and is not reflected on the map.

The sickle cell projects conducted 222 awareness activities during SFY 2018. These activities were estimated to have made 15,155,389 contacts through a variety of modes. In Figure 12, community or professional outreach engagement included walks, health fairs, and exhibits. Social media engagement included outlets such as Facebook and Twitter, while traditional media engagement included newspaper/print, television, radio, and electronic newsletter awareness activities.

Figure 12. Awareness Activities by Mode (n=222)



Source: Sickle Cell Projects- Awareness/Media Activity Reporting SFY 2018

In September 2018, each of the seven ODH-funded sickle cell projects conducted awareness activities during Sickle Cell Sabbath. Sickle Cell Sabbath is an interfaith outreach campaign designed to educate and increase awareness within the faith community about SCD and the need for minority blood donors. Projects reported reaching 365 faith-based organizations in the state. Sickle Cell Sabbath activities were estimated to have made 25,525 contacts.

Source: Sickle Cell Projects- Awareness/ Media Activity Reporting SFY 2018





Appendices

APPENDIX A: Sickle Cell Facts

Sickle cell disease (SCD) is a term used to describe a group of inherited disorders characterized by the predominance of hemoglobin-S in the red blood cells. These inherited disorders include:

- **Sickle cell anemia (HbSS)**
People who have this form of SCD inherit a gene for sickle hemoglobin (HbS) from each parent. HbSS is the most common type of SCD and is usually the most severe form of the disease.
- **Hemoglobin sickle C disease (HbSC)**
People who have this form of SCD inherit a gene for HbS from one parent and from the other parent a gene for an abnormal hemoglobin called “C.” Hemoglobin is a protein that allows red blood cells to carry oxygen to all parts of the body. HbSC is usually a milder form of SCD.
- **Hemoglobin sickle beta (β) thalassemia syndromes (HbS β^+ and HbS β^0 -thalassemia)**
People who have this form of SCD inherit one gene for HbS from one parent and one gene for β thalassemia, another type of anemia, from the other parent. There are two types of β thalassemia: HbS β^+ -thalassemia and HbS β^0 -thalassemia. Those with HbS β^0 -thalassemia usually have a clinical course similar to HbSS. People with HbS β^+ -thalassemia tend to have a milder form of SCD.
- **Hemoglobin SD, Hemoglobin SE, and Hemoglobin SO (HbSD, HbSE and HbSO)**
People who have these rare forms of SCD inherit one gene for HbS and one gene from an abnormal type of hemoglobin (“D,” “E,” or “O”). The severity of these rarer types of SCD varies.

Sickle cell trait (SCT) is the “**carrier**” state in which a person inherits one gene for HbS from one parent and one normal gene (“A”) from the other parent. Most people with SCT usually do not have any of the symptoms of SCD but can pass the trait on to their children.

According to the [Centers for Disease Control and Prevention](#), the exact number of people living with SCD is unknown. In the United States, it is estimated that:

- SCD affects approximately **100,000 Americans**.
- SCD occurs among about **1 out of 365** Black or African-American births.
- SCD occurs among about **1 out every 16,300** Hispanic-American births.
- **About 1 in 13** Black or African-American babies are born with SCT. ⁴

SCD is one of the most commonly inherited diseases worldwide. While most predominant among Blacks or African-Americans in the U.S., individuals with heritage from sub-Saharan Africa; Spanish-speaking regions in the Western Hemisphere (South America, the Caribbean, and Central America); Saudi Arabia; India; and Mediterranean countries such as Turkey, Greece, and Italy may also be affected. Globally, it is estimated that:

- SCD occurs in approximately **300,000 births** annually.
- **SCD is most prevalent in malaria endemic parts of the world**, primarily Africa, the Middle East, and South Asia.
- In many African countries **10% to 40%** of the population carries the sickle-cell gene, resulting in estimated SCD prevalence of at least 2%.⁵

SCD, which causes a wide range of severe and even life-threatening consequences, is caused by a mutation in the DNA instructions for hemoglobin, a protein vital for carrying oxygen in the blood. As a result of this mutation, individuals with SCD experience lifelong complications including anemia, infections, stroke, tissue damage, organ failure, intense painful episodes, and premature death. These debilitating symptoms and the complex treatment needs of individuals living with SCD often limit their education, career opportunities, and quality of life.⁵

APPENDIX B: Sickle Cell Project Profile - Region I

Cincinnati Comprehensive Sickle Cell Center

Cincinnati Children's Hospital Medical Center

3333 Burnet Ave., MLC 7015

Cincinnati, OH 45229

(513) 636-7541

<https://www.cincinnatichildrens.org/service/s/sickle-cell>

Region I Counties:

Adams, Brown, Butler, Clermont, Clinton, Hamilton, Highland, Warren

ODH SFY 2018 Funding Allocation: \$125, 957

Standard Services and Programs:

- Newborn screening coordination, counseling, and education.
- Outreach education and training for health care professionals and the public.
- Care coordination for pediatric patients diagnosed with sickle cell disease.
- Regional center for hemoglobinopathy educational materials.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) educational activities.

Expanded Services and Programs:

- Annual newsletter for regional health care providers.
- "Center Talk" newsletter for pediatric families.

Non-Grant Supported Services and Programs provided through the Comprehensive Sickle Cell Center at Cincinnati Children's Hospital Medical Center:

- Comprehensive sickle cell clinic for pediatric patients, including social work and school intervention program.
- Tailored educational and support programs for individuals/families with sickle cell disease.
- Extensive clinical research studies for individuals/families with sickle cell disease.
- Transition to adult care program with University of Cincinnati Adult Sickle Cell Clinic.
- Quality improvement outcomes projects for sickle cell disease, including a regional six-state learning network (Sickle Treatment Outcomes Research in the Midwest – STORM).
- Annual summer camp program.
- Health care provider education programs including the annual National Hemoglobinopathy Counselor Training Course.
- Training/rotation in hemoglobin disorders.

Project Director: Lisa Shook, MA, MCHES

Medical Director: Charles Quinn, MD

APPENDIX C: Sickle Cell Project Profile - Region II

West Central Ohio Comprehensive Sickle Cell Center

Dayton Children's Hospital

One Children's Plaza

Dayton, OH 45404

(937) 641-3111 or 1-800-228-4055 ext. 5014

<https://www.childrensdayton.org/patients-visitors/services/hematology-oncology/sickle-cell-disease>

Region II Counties:

Allen, Auglaize, Champaign, Clark, Darke, Greene, Hancock, Hardin, Logan, Mercer, Miami, Montgomery, Paulding, Preble, Putnam, Shelby, Van Wert

ODH SFY 2018 Funding Allocation: \$72,932

Standard Services and Programs:

- Newborn screening coordination and follow-up.
- Free hemoglobin testing for adults of child-bearing age.
- Hemoglobinopathy counseling and education.
- Care coordination and specialty resource referrals.
- Community education and outreach.
- Regional resource clearinghouse of educational materials.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) activities.

Expanded Services and Programs:

- Patient and family disease education.
- School intervention program.
- Patient assistance program.
- Special family events/activities.
- Outreach clinics at Specialty Care Center – Lima (St. Rita Eastside Urgent Care Center, 967 Bellefontaine Ave., Lima) and South Campus – Springboro (3333 W. Tech Road, Miamisburg).

Non-Grant Supported Services and Programs provided through the Hematology/Oncology

Department at Dayton Children's Hospital:

- Diagnostic evaluation/treatment/management services for patient birth to 22 years of age.
- Consultation and collaboration with adult health care providers.
- Nutrition assessment and counseling.
- Social work services.
- Psychosocial assessment and intervention.
- Transition Clinic at Five Rivers Health Center (725 South Ludlow St., Dayton).
- Training/rotation/internship in hemoglobin disorders.

Project Director: Cynthia L. Moon, MSE

Medical Director: Mukund Dole, MD

APPENDIX D: Sickle Cell Project Profile - Region III

Sickle Cell Project of Northwest Ohio

313 Jefferson Ave. (mailing address)
Toledo, OH 43604

Nexus (location address)

1415 Jefferson Ave.

Toledo, OH 43604

(419) 214-5700, ext. 6240

<https://nhainc.org/sickle-cell-project-nwo/>

Region III Counties:

Defiance, Erie, Fulton, Henry, Huron, Lucas, Ottawa, Sandusky, Seneca, Williams, Wood

ODH 2018 Funding Allocation: \$54,743

Standard Services and Programs:

- Newborn screening coordination and follow-up services.
- Hemoglobinopathy counseling (off-site locations at Toledo Children's Hospital and Mercy Children's Hospital).
- Education, outreach, and awareness activities in 11 counties.
- Regional resource center for hemoglobinopathy educational materials.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) activities.

Expanded Services and Programs:

- Patient/family disease education (Toledo's Children's Hospital and Mercy Children's Hospital).
- School outreach and interventions (including post-secondary collaboration with disability service programs).
- Genetic counseling (in collaboration with referral to the University of Toledo Division of Genetics).

Non-Grant Supported Services and Programs provided through Nexus:

- Primary and pediatric medical care for uninsured and underinsured populations.
- Social work services.
- On-site pharmacy and laboratory.
- WIC Services.
- Dental Services.
- Women's Health.
- Mildred Bayer Homeless Clinic.

Project Director: La'Shardae Scott

Medical Advisors: Crawford Strunk, MD (Toledo Children's Hospital) and Melisa Mullins, MD (Mercy Children's Hospital)

APPENDIX E: Sickle Cell Project Profile - Region IV

Comprehensive Sickle Cell Disease and Thalassemia Program

Nationwide Children's

700 Children's Drive

Columbus, OH 43205

(614) 722-5948

<http://www.nationwidechildrens.org/sickle-cell-and-thalassemia-program>

Region IV Counties:

Athens, Belmont, Coshocton, Delaware, Fairfield, Fayette, Franklin, Gallia, Guernsey, Harrison, Hocking, Jackson, Jefferson, Knox, Lawrence, Licking, Madison, Marion, Meigs, Monroe, Morgan, Morrow, Muskingum, Noble, Perry, Pickaway, Pike, Ross, Scioto, Union, Vinton, Washington, Wyandot

ODH 2018 Funding Allocation: \$171,595

Standard Services and Programs:

- Newborn screening coordination and follow-up services.
- Hemoglobinopathy counseling.
- Hemoglobinopathy education, outreach, and awareness activities.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) activities.

Expanded Services and Programs:

- School/daycare intervention program.
- Patient/family disease education.
- Blood/bone marrow donor education and registry drives.
- Nursing education.
- Transition to adult care.
- Satellite clinic at Nationwide Children's Hospital – Close to Home with Urgent Care Center (6435 E. Broad St., Columbus).

Non-Gant Supported Services and Programs provided through the Hematology/Oncology/BMT Division at Nationwide Children's:

- Infusion services.
- Confirmatory testing for abnormal newborn screening hemoglobin results.
- Comprehensive medical management and follow-up for hemoglobin disease.
- Genetic counseling and extended family testing.
- Case management.
- Dental evaluations.
- Psychological testing and evaluations.
- Apheresis Program.
- Psychological/clinical research trials.
- Student training.

Project Director: Tanica Jeffries, MS, LPC, LSW

Medical Director: Anthony Villella, MD

APPENDIX F: Sickle Cell Project Profile - Region V

American Sickle Cell Anemia Association

10900 Carnegie Ave.

DD Building, Suite DD1-201

Cleveland, OH 44106

(216) 229-8600

<http://www.ascaa.org>

Region V Counties:

Cuyahoga, Geauga, Lake, Lorain/Elyria, Medina

ODH 2018 Funding Allocation: \$188,164

Standard Services and Programs:

- Newborn screening follow-up and diagnostic testing.
- Supportive services/crisis intervention/community referrals for individuals and families.
- Hemoglobinopathy counseling and education.
- Collaboration and direct services to maternity hospitals and birthing centers in Region V.
- Resource center for hemoglobinopathy education materials.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) activities.

Expanded Services and Programs:

- Hispanic community direct services and outreach.
- Global/international education and referral initiative.
- School/home intervention program.
- Collaborative relationships with pediatric and adult hospitals in Region V.
- CBS Cares public service announcement for sickle cell disease (local and national airing).
- Website (in over 100 pull down languages).
- Facebook, Twitter, YouTube, and Instagram.
- "Sickle Cell" online media series.

Non-Grant Supported Services and Programs provided through the American Sickle Cell Anemia Association:

- On-site diagnostic hemoglobin testing for the general public (in association with the Cleveland Clinic).
- Summer Cleveland Clinic Science Internship Program for high school students.
- Cleveland Clinic resident physician rotation program.
- Bryant & Stratton Pre-Graduate Internship Program (Science and Administration).

Project Director: Ira Bragg-Grant

Medical Advisor: Grace Onimoe, MD (MetroHealth Hospital)

APPENDIX G: Sickle Cell Project Profile - Region VI

Ohio Region VI Sickle Cell Program

Akron Children's Hospital

One Perkins Square

Akron, OH 44306

(330) 543-3521 or (800) 262-0333, Ext. 8730

<https://www.akronchildrens.org/departments/Sickle-Cell-Program.html>

Region VI Counties:

Ashland, Ashtabula, Carroll, Columbiana, Crawford, Holmes, Portage, Mahoning, Richland, Stark, Summit, Trumbull, Tuscarawas, Wayne

ODH SFY 2018 Funding Allocation: \$96,609

Standard Services and Programs:

- Newborn screening coordination and follow-up.
- Patient and family advocacy.
- Hemoglobinopathy counseling and patient/family education.
- Resource center for audiovisual and print materials regarding hemoglobinopathies.
- Physician communication on newborn screening and follow-up standards and practices.
- Hemoglobinopathy education, outreach, and awareness activities for health care professionals/providers, consumers, and the public.
- Sickle Cell Sabbath/Sunday and Sickle Cell Awareness Month (September) activities.

Expanded Services and Programs:

- School intervention services.
- Quarterly newsletter.
- Patient and family disease education.
- Adolescent transition program.
- Holiday events for patients and their families.
- Monthly parent/patient support group meetings.
- Psychosocial assessments/interventions and supportive services.
- Annual sickle cell awareness walk.
- Annual weekend summer camp, **Camp Ed Bear** (Children's Hematology-Oncology Patients and Staff), for patients ages 6-16 and Counselor in Training Program for patients age 16-21.

Non-Grant Supported Services and Programs provided through the Showers Family Center for Childhood Cancer and Blood Disorders at Akron Children's Hospital:

- Monthly comprehensive clinics for sickle cell disease patients (with incorporated genetic, psychosocial counseling, nutrition, physical therapy, and radiology services).
- Confirmatory testing for abnormal newborn screening hemoglobin results.
- Individual hematological consultation and diagnostic testing.

Project Director: LaTonya Lewis

Medical Director: Prasad Bodas, MD

APPENDIX H: Sickle Cell Project Profile - Statewide

Ohio Sickle Cell and Health Association, Inc

341 South Third St.

Suite 200

Columbus, OH 43215

(614) 228-0157

<http://www.ohiosicklecell.org>

Counties: Statewide-88 Ohio Counties

SFY 2018 Funding Allocation: \$90,000

Standard Services and Programs:

- Consumer and professional training and education.
- Statewide public awareness and media campaigns.
- Sickle Cell Sabbath/Sunday and Sickle Cell Month (September) activities.

Expanded Services and Programs:

- Empowerment Scholarship Fund for educational events.
- Empowerment events for young adults/adults living with sickle cell disease.
- Social media engagement and marketing of statewide and regional sickle cell activities and events.
- Advisory organization to the Ohio Sickle Cell Affected Families Association.
- Ohio Chapter of the National Organization - Sickle Cell Disease Association of America (SCDAA).

Non-Grant Supported Services and Programs (provided through the Ohio Sickle Cell and Health Association and Sickle Cell Disease Association of America):

- Case management and physician referral (per request).
- Patient/client/legislative advocacy for issues related to sickle cell disease.
- Information and referral services for extended family testing.
- Emergency support services for individuals/families with sickle cell disease.
- Social marketing campaigns for individuals/families with sickle cell disease.
- Statewide coordination of Sickle Cell Runs, Walks and Jogs in Ohio.
- Statewide collaboration and coordination of adult (including young adult) transition services.
- "Get-Connected" SCDAA database collection.
- CBO training and development.
- Community Health Worker Training.

Project Director: Annie J. Ross-Womack

Medical Advisor: Payal Desi, MD (The Ohio State University Comprehensive Sickle Cell Clinic)

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3. Ohio population by County-percentage of Black residents. Available at <https://us-places.com/Ohio/black-percentage-population-comparison.htm>.

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