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RuSH Strategies from the Field: Data Collection

Georgia Health Policy Center

Registry and Surveillance System for Hemoglobinopathies

RuSH

Strategies from the Field: Data Collection

DEPARTMENT OF HEALTH & HUMAN SERVICES · USA

CDC
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National Heart Lung and Blood Institute

Registry and Surveillance System for Hemoglobinopathies

RuSH

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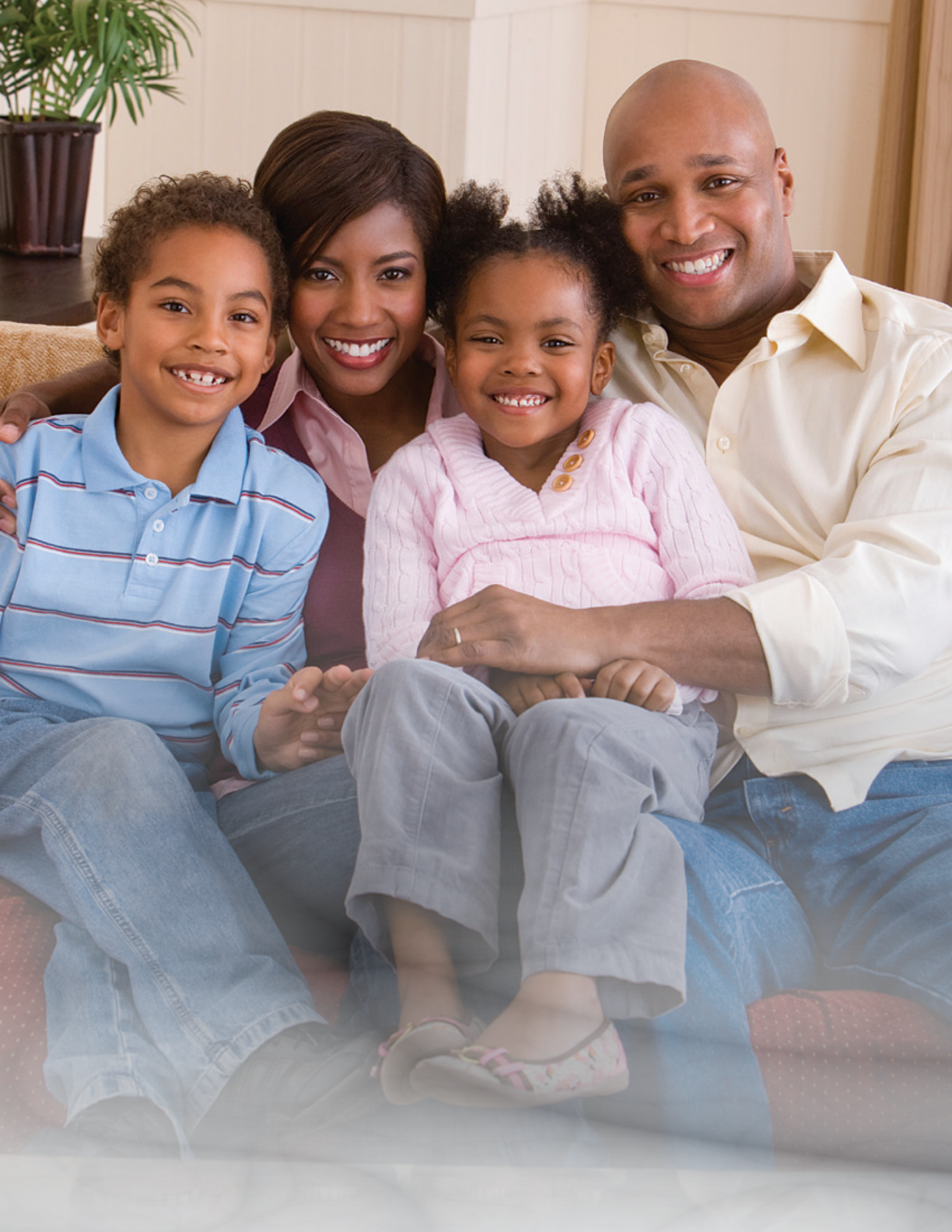


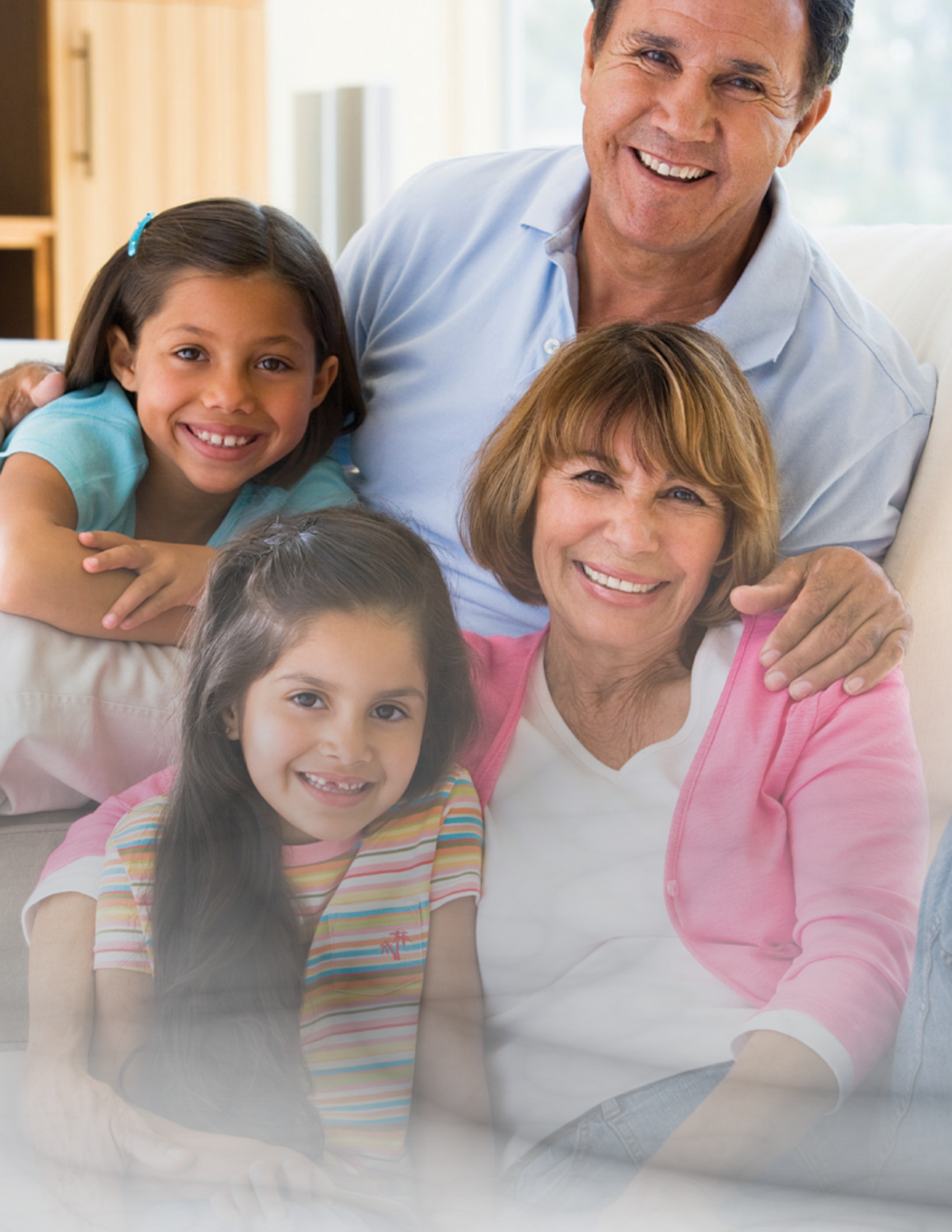
Overview

In 2010, the Registry and Surveillance System for Hemoglobinopathies (RuSH) pilot project was implemented by the Centers for Disease Control and Prevention (CDC) to collect state-specific, population-based data on people with sickle cell disease (SCD) and thalassemia. The 2-year pilot project was supported and conducted in collaboration with the National Institutes of Health's National Heart, Lung, and Blood Institute (NHLBI).

Overall project goals included determining the number of people with SCD and thalassemia and increasing knowledge and awareness about health care use and outcomes. Seven states were funded to participate in data collection: California, Florida, Georgia, Michigan, New York, North Carolina, and Pennsylvania. Due to the unique nature of the available data sources in each state, a variety of data collection methods were employed.

It is anticipated that these state-based surveillance data will:

- Provide estimates of the number of new cases (incidence) of SCD and thalassemia each year.
- Provide estimates of the total number of existing cases (prevalence) of SCD and thalassemia.
- Provide information on trends in medical care for people with these disorders.
- Provide information about complications and death rates.
- Provide information to assist with planning public health interventions (i.e., services, health promotion campaigns, health education, and training) to improve the health of these populations.



Strategy Field Reports

The purpose of this document is to showcase the unique ways in which the states implemented activities designed to meet the programmatic goals of RuSH. In the following sections, each state highlights a particular aspect of its data collection process, including information about the resources needed to operate or replicate the activity and lessons learned. The intent is to give a brief overview of the complexity of this project and to provide contact and programmatic information to other entities for the potential modification or replication, or both, of activities.

The information in this report was provided by programmatic team members who worked with CDC staff in spring 2012.

Case Definition

An important initial step in developing the RuSH pilot project was defining the patients with hemoglobinopathies that were included in the surveillance efforts. Because most of the collected information came from preexisting data sources, and these data sources varied greatly among the seven states, a case definition was created to help standardize the studied population as much as possible. A workgroup consisting of clinicians and public health professionals was established to help CDC and the NHLBI in creating the definition. Please note that this definition and its corresponding complications, procedures, and treatments will be referred to throughout this document.

LEVEL 1: HIGH

- Results of a state newborn screening program with confirmatory testing, or
- Clinical diagnosis by a physician with documented confirmatory laboratory testing after the newborn period.

LEVEL 2: PROBABLE

- Results of a state newborn screening program without confirmatory testing, or
- Hemoglobinopathy-related International Classification of Disease (ICD) -9 or -10 code (see following codes; excluding sickle cell trait) used during two or more separate health care encounters, plus one or more hemoglobinopathy-associated complication, treatment, or procedure (see following lists)

LEVEL 3: POSSIBLE

- Sickle cell trait ICD -9 or ICD-10 code (see following codes) used during two or more separate health care encounters, plus one or more hemoglobinopathy-associated complication, treatment, or procedure (see following lists), or
- Single health care encounter with a hemoglobinopathy-related ICD -9 or ICD-10 code(see following codes; excluding sickle cell trait)

*Health care encounter: Inpatient or outpatient hospitalization or health care visit, including emergency department visits.

Hemoglobinopathy-Related International Classification of Disease (ICD) Codes

ICD-9 Codes

282.4 Thalassemias

282.41 Sickle cell thalassemia without crisis

282.42 Sickle cell thalassemia with crisis

282.49 Other thalassemia

282.6 Sickle cell disease

282.5 Sickle cell trait

282.60 Sickle cell disease, unspecified

282.61 Hb-SS disease without crisis

282.62 Hb-SS disease with crisis

282.63 Sickle cell/Hb-C disease without crisis

282.64 Sickle cell/HB-C disease with crisis

282.68 Other sickle cell disease without crisis

282.69 Other sickle cell disease with crisis

ICD-10 Codes

D56 Thalassemia

D56.0 Alpha thalassemia

D56.1 Beta thalassemia

D56.2 Delta-beta thalassemia

D56.8 Other thalassemias

D56.9 Thalassemia, unspecified

D57 Sickle cell disorders

D57.0 Sickle cell anemia with crisis

D57.1 Sickle cell anemia without crisis

D57.2 Double heterozygous sickling disorders

D57.3 Sickle cell trait

D57.8 Other sickle cell disorders

Hemoglobinopathy-Associated Complications

1. Chronic renal failure/Proteinuria
2. Pneumonia, acute chest syndrome
3. Pulmonary hypertension
4. Stroke (ischemic or hemorrhagic), transient ischemic attack, seizures
5. Intracranial bleeding
6. Priapism
7. Iron overload
8. Gallstones/cholelithiasis, cholecystitis
9. Avascular necrosis
10. Retinal disease
11. Splenomegaly, splenic sequestration, hypersplenism
12. Leg ulcers
13. Dactylitis
14. Osteomyelitis

Hemoglobinopathy-Associated Treatments

1. Hydroxyurea
2. Parenteral analgesics
3. Iron Chelators
4. Erythropoietin
5. Folic acid

Hemoglobinopathy-Associated Procedures

1. Red cell transfusion
2. Red cell exchange
3. Splenectomy
4. Cholecystectomy
5. Transcranial Doppler

North Carolina: Matching Existing Government and Clinical Datasets

Program Overview

The goal of the North Carolina (NC) RuSH project was to identify the number of people with sickle cell disease (SCD) or thalassemia who received care and services in North Carolina during the years 2004–2008. A secondary goal was to determine what health services this population received during the same timeframe. Specifically, the NC RuSH project was designed to collect information from specific datasets from various governmental and clinical data sources. The existing datasets included information on demographics, clinical diagnoses (i.e., laboratory test or International Classification of Disease, 9th Revision or 10th Revision), mortality, health care use, medical complications, procedures, and treatment modalities.

This surveillance effort supports North Carolina's commitment to ensure that people living with SCD or thalassemia lead an independent, healthy life. Moreover, the gathered surveillance data will guide the health practices for program partners and health care professionals, to better serve populations affected by hemoglobinopathies. Ultimately, this will increase the knowledge and awareness of SCD and thalassemia for stakeholders, policy makers, and residents of North Carolina. In addition, it will ensure the best use of allocated resources for the North Carolina Division of Public Health's Sickle Cell Syndrome Program (NCSCSP) and better streamline educational and outreach strategies.

Resources Needed:

To successfully conduct statewide surveillance of people with SCD or thalassemia, it was critical to determine the systems within state government that provided access to information on the targeted populations and attain legal access to these systems. In particular, letters of agreement or memorandums of agreement, or both, were established with various governmental agencies, clinical partners, and community entities to capture patient data. Following is a list of agencies with which agreements were established to facilitate the project:

6. North Carolina Women's and Children's Section Web-based System (WCSWeb)—NCSCSP client data
7. North Carolina State Center for Health Statistics—hospital discharge, Medicaid, and birth and death records
8. North Carolina Disease Event Tracking and Epidemiologic Collection Tool (NC DETECT)—emergency department data
9. North Carolina Purchase of Medical Care Program (POMC)—state program insurance claims data
10. North Carolina Immunization Registry (NCIR)—vaccination data
11. North Carolina State Laboratory of Public Health (SLPH—newborn and nonnewborn data
12. North Carolina Comprehensive Medical Centers—six comprehensive sickle cell centers' client data
13. University of North Carolina at Greensboro—data analysis and evaluation

Other resources included staff with specific skill sets, such as information technology (IT), programmers/coders, data managers, program coordinators, and epidemiologists. Time required for data coordination was substantial, as 75%–100% of the time of one FTE was needed. There also needed to be consideration for the availability of appropriate computer software and hardware such as SAS, SPSS, and Microsoft® Excel 2010 to extract, deduplicate, and link data. Budget cost can be significant if the skill sets and other technology are not readily available. It is advisable to use in-kind services when possible to offset cost. These resources were essential for matching existing datasets within surveillance systems.

Lessons Learned:

Goals and objectives need to be clearly defined at the onset of the project; changes in these can affect the project and cause delays, as well as increase cost and resources. There must be adequate time allowed so that all partners are able to agree on deliverables and clearly define the expectations for the work. If competing agendas and issues of data privacy arise, these may challenge the prioritization of surveillance activities. There is a

chance that substandard data may be delivered, so this will require additional review and analysis. Validation of the quality of the data is necessary. Finally, if only limited information is available from the datasets requested from participating partners, this may inhibit the matching of data among data sources.

Outcome:

Preliminary findings identified approximately 8,955 unique individuals with sickle cell disease or thalassemia who were living in North Carolina during the years of 2004–2008. Roughly 50% were Level 1 cases, 14% were Level 2 cases, and 36% were Level 3 cases. Analysis of these data allowed public health officials to derive prevalence and incidence estimates of hemoglobinopathies in North Carolina, as well as the effects of these diseases.

Data collection for this activity provided opportunities in the enhancement and evaluation of the NCSCSP. It benefitted patients, health care providers, community-based organizations, faith-based communities, policy makers, and the general public. This type of information helped to determine quality of care standards, service delivery, and resource distribution. In addition, it created a road map and model for accessing data and building interagency collaboration within state government. The final analysis of the data provided information and best practices to:

1. Outline a clear plan and approach for improved medical treatment, services, and resources.
2. Make a case for continued support of the program.
3. Direct efforts to targeted populations with the greatest need.
4. Shape the development of health promotion messages and outreach strategies.
5. Increase knowledge of general public.
6. Track trends of the effects of these conditions on individuals throughout their lifespan.

Sustainability/Translation:

The NCSCSP is committed to advancements in the treatment, research, legislation, and quality of care for people with SCD or thalassemia, which ultimately might lead to a cure for these diseases. The RuSH project laid a foundation and framework to provide a global platform to address hemoglobinopathies at a level that examines the diseases beyond diagnosis and treatment. The NC RuSH effort will continue to advocate and seek resources and opportunities to build a registry for hemoglobinopathies. Efforts that will continue beyond RuSH funding include sharing of interagency data, enhancement of SLPH with the WCSWeb and building of statewide partnerships.

The surveillance for hemoglobinopathies provides a model to investigate the trends and epidemiology of other health conditions as well, such as heart disease, stroke, or Alzheimer disease. The NC RuSH project could serve as a model for all areas of health care to ensure that work being done is adequate to address the needs of the community affected by hemoglobinopathies and is based on sound data and practices. This work is translatable to other areas of public health because it sets a standard and is an impetus for surveillance. Surveillance for hemoglobinopathies should be a basic public health activity to ensure that the appropriate protocols, treatments, and prevention efforts are being provided at the federal, state, and local levels.

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Georgia: Limitations of International Classification of Disease (ICD) Codes for Defining and Identifying Hemoglobinopathy Cases

Program Overview

Each of the RuSH states used International Classification of Disease (ICD) codes to identify people with hemoglobinopathies in at least one administrative dataset. States also used the ICD ninth or tenth revision (ICD-9 or ICD-10) codes to classify mortality data from death certificates and the International Classification of Diseases, Clinical Modification (ICD-9-CM or ICD-10-CM) codes to classify morbidity data from inpatient and outpatient records, public and private health insurance claims, and public health programs. While several subcategories of ICD codes were available to distinguish between different types of hemoglobinopathies, the coding for clinically relevant types of hemoglobinopathies in administrative datasets might not always be specific. Therefore, ICD codes for common comorbid conditions were sought in hospital discharge and billing data from public and private insurance programs and used to add specificity to case classification, along with Current Procedural Terminology® (CPT®) and National Drug Codes (NDC) for common medical procedures and treatments found in outpatient and pharmacy billing data.

Resources Needed

Identification of hemoglobinopathy cases in Georgia was a team effort, as much of the administrative data necessary for the surveillance project were not within the purview of Public Health. Thus, the project required partnerships and data-sharing agreements between Public Health and the state Medicaid/Children's Health Insurance Program (CHIP) agency, as well as the entity that collects, standardizes, and manages the state's hospital discharge data, the Georgia Hospital Association (GHA), a nonprofit trade organization. It was necessary for Public Health and the Georgia RuSH team to acquire identified, person-level data from these partners. In return, partners were assured that the data would be secured and the process would adhere to all Health Insurance Portability and Accountability Act protections. Georgia also had a benefit in this effort, as both sickle cell disease (SCD) and thalassemia are reportable conditions for children up to 18 years of age through surveillance for birth defects. Mining, coding, and deduplicating each of the datasets required the expertise of at least a part-time data analyst comfortable working with large administrative datasets. In addition to the data analyst, the principal investigator and Public Health representative also devoted significant personnel time to securing necessary data-sharing agreements.

Lessons Learned

Administrative data are collected for billing purposes, not surveillance or research. Many individual records had several ICD codes listed because the provider considered them as possible diagnoses, even if those diagnoses later were ruled out. Therefore, it was important to look for more than one instance of a particular diagnosis when attempting to count true cases of disease.

Case comparison across datasets was limited due to the lack of consistency in ICD coding for the hemoglobinopathy-related complications, treatments, and procedures used in the RuSH case definition. For example, while hospital discharge and billing data provided evidence for complications via ICD codes, only outpatient billing data supplied CPT® and NDC to identify medical procedures and treatments.

ICD codes were better at identifying SCD than thalassemia because there were codes for subcategories of SCD, including a code for sickle cell trait. There was only one category for thalassemia in the ICD-9 classification and it did not distinguish between thalassemia trait and thalassemia disease.

It was possible to create a surveillance system with administrative data and it was determined that datasets with better unique patient identifying information were more favorable for matching and deduplication purposes. In the absence of unique identifiers like social security numbers, many assumptions needed to be made and deduplication took a significant investment of time.

Outcome

The Georgia RuSH team was able to use the individual-level hospital discharge data to show the frequency of emergency department (ED) visits and inpatient stays in each county, and to characterize disease prevalence by age group, sex, race and ethnicity, and county of residence. This information was provided in poster form to sickle cell support groups around the state for use in their outreach and advocacy efforts. Findings reinforced community experience regarding areas of the state with the greatest disease burden, the needs of youth transitioning to adult care, and the nature of hospital spending on hemoglobinopathy-related care. Better prevalence data now can be shared with partners. For example, the percentage of patients with SCD covered by Medicaid can be estimated. Medicaid claims data also can be used to evaluate the frequency and type of care that patients with hemoglobinopathies receive to help target provider education efforts.

Sustainability/Translation

Additional funding is necessary to design and complete validation studies that determine the accuracy of the RuSH case definition. That is, what percentages of Level 2 and Level 3 cases actually have a hemoglobinopathy? Without additional funding, it also will not be possible to define and identify additional hemoglobinopathy cases outside of the study period. One sustainable activity will be the use of administrative data to advance the knowledge of health service use by individuals with hemoglobinopathies.

This project can help to inform surveillance activities for other noninfectious, chronic conditions that affect individuals throughout their lifetime. In Georgia, a first step would be to approach the GHA and state Medicaid agency about reporting all birth defects of children up to 8 years of age. While birth defects currently are reportable in Georgia, the system is passive and reporting compliance is low. Insurance or provider organizations, or both, could provide a service for their members by periodically searching their administrative databases for specific ICD codes and reporting cases to the Public Health Department.

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California: Obtaining and Using Medicaid Data for Hemoglobinopathy Surveillance

Program Overview

California's focus in the RuSH project has been to identify as many cases as possible of sickle cell disease (SCD) and thalassemia using administrative and clinical data. Using the methodology and case definitions developed by the seven funded states and the Centers for Disease Control and Prevention, over 5,800 confirmed or probable cases of SCD were identified in the state in 2008, which represents over 90% of the estimated number of cases based on extrapolations of case count from demographic data.[1] Additionally, over 1,000 confirmed cases of alpha or beta thalassemia were identified in 2008, and algorithms are being developed to better determine probable cases from the data.

Obtaining administrative and clinical data to do this work was successful. Access was granted to (1) newborn screening data going back as far as 1999; (2) all electronically available hospital discharge and emergency room visit data and vital records (birth and death certificates); and (3) data for patients with SCD or thalassemia attending clinics at the Children's Hospital & Research Center Oakland and Children's Hospital Los Angeles (both have adult SCD and thalassemia clinics, as well as pediatric clinics). In addition, Medi-Cal (California's Medicaid system) data related to members with one or more SCD- or thalassemia-related International Classification of Disease, (ICD-9) codes was made available.

Medi-Cal data were the most difficult data to obtain and to work with; however, while each of these sources was vital in helping identify cases or contributing key information about cases found, or both, Medi-Cal data contained the greatest number of cases not seen elsewhere and the most information about cases seen.

Resources Needed

California RuSH employed two full-time staff: a project director/epidemiologist responsible for obtaining data from agencies and clinics and a database developer who managed and cleaned all datasets, developed matching algorithms across them, and linked and deduplicated the cleaned data. California's Department of Health Care Services (DHCS) is the agency responsible for the Medi-Cal program, and data were requested directly from it for this project. The state's Committee for the Protection of Human Subjects declared the RuSH project to be a nonresearch surveillance activity prior to the contract being established with the DHCS; however, the DHCS required a review and approval by its own Data and Research Committee before granting access to data. This review was a multistep process in which the RuSH project staff requested particular variables, such as social security numbers, patient ZIP Codes, and exact dates of birth from all claims related to individuals who had one or more SCD- or thalassemia-related ICD-9 codes during the period 2000–2008. The committee approved, denied, or requested more information on the justification for each variable. Approval took over 6 months, while transfer of the initial data set took an additional 4 months from approval to receipt.

Once the initial dataset was received from the DHCS, it was compared to the datasets already received from other agencies and clinic partners, and a list of cases seen in the other sources—but not in the Medi-Cal data—was created. That list was sent to the DHCS with a request to pull all claims seen for those individuals as well; this request was granted quickly. Once all data were received, weighted matching algorithms were developed to enable linking and deduplication of data using social security numbers. When linking had taken place, work with the claims themselves was begun. Because this data source had not been developed with an eye toward research or surveillance (as in the case of hospital discharge

data, for example), extensive data cleaning and translation were necessary. Five different billing code systems were used (e.g., International Classification of Disease, 9th Revision; Current Procedural Terminology®; and Healthcare Common Procedure Coding System) in one variable, and these needed to be standardized. Some identified cases had two distinct identification numbers: some claims were located by social security number, while others were found using a county-specific Medi-Cal identifier. Rather than receiving cleaned data, the RuSH team received a data file that included all claim "lines"; because of this, many of the encounters in the claims data were entered multiple times to account for claim corrections and resubmissions. These duplicate entries were accounted for in analyses involving counts of visits or claims or dollar amounts. Solving these problems was one of the most time-consuming elements of the RuSH project.

Lessons Learned

States using Medicaid data for disease surveillance would be advised to recognize the differences between that data source and other administrative data sources. For example, hospital discharge data produced at the state level can be a research- or surveillance-ready product, with variables cleaned and well explained in a data dictionary. Budgeting extra time for obtaining and working with the data is advised. Additionally, should an "eligibility file" describing when members' eligibility for benefits began and ended be available, states are advised to request that information. Such files allow the analysis of Medicaid members as a cohort with appropriate censoring dates.

Outcome

Despite challenges, Medi-Cal data were found to be well worth the time and effort to the project. In the case of SCD, an estimated 545 (14%) of 3,901 probable cases were identifiable only through the Medi-Cal record, a much greater proportion than from any other single data source, and 3,958 (64%) of 6,208 confirmed and probable SCD cases had data in the Medi-Cal file. Similarly, among the 1,920 probable cases of thalassemia, 287 (15%) were seen only in the Medi-Cal data, and 6,194 (38%) of the 16,504 probable or possible cases were seen in the Medi-Cal data. Additionally, all information about outpatient services and prescription treatments for identified cases came from this source.

Sustainability/Translation

California currently is seeking other funding sources to continue surveillance of hemoglobinopathies and looking toward the development of an ongoing patient registry. While there is no funding in place to continue the collection of administrative or clinical data, the CA RuSH team is optimistic that the utility of the work done to understand the public health implications of hemoglobin disorders among the state's population will lead to funding for continued work. In addition to establishing a baseline count and understanding of the effects of SCD and thalassemia among the population, this work has set the stage for the use of administrative and clinical data to conduct surveillance for other genetic disorders.

The use of Medicaid claims data in conjunction with other data sources can be a powerful tool in public health surveillance, allowing both effective case identification and an abundance of information on complications, treatments, and outcomes for the cases, including outpatient and prescription information not readily available via other administrative data sources. Obtaining and working with the data were not straightforward, but the data's utility more than made up for the difficulties in gaining access to it.

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Pennsylvania: The Use of Partnerships in Creating a Hemoglobinopathy Database

Program Overview

The Pennsylvania Department of Health (PA DOH) developed a linked data surveillance system for patients with sickle cell disease (SCD) or thalassemia in Pennsylvania. Due to privacy issues that could not be resolved within a reasonable time frame, the PA DOH could not obtain data from vital statistics, Medicaid, or the Pennsylvania Health Care Cost Containment Council's (PHC4) hospital discharge database. As a result, the PA DOH established a partnership with the three leading childrens hospitals in Pennsylvania: St. Christopher's Hospital for Children, The Children's Hospital of Philadelphia® (CHOP), and Children's Hospital of Pittsburgh (CHIP) to aggregate descriptive data about their respective populations with SCD and thalassemia, as well as Carnegie Mellon University (CMU) to provide analytical support for de-identifying, cleaning, and linking the databases. The databases used to link the newborn screening dataset to the vital statistics data were the hospital administrative databases from the previously listed hospitals. These databases were created to incorporate data specific to patients with SCD or thalassemia. These datasets contained the data elements that were decided on by the RuSH Data and Harmonization workgroup. Fine-grained Record Linkage software was used to link all of the databases used in this project. The individuals on the RuSH team from each of the three childrens hospitals negotiated with the information technology department at their respective facilities to create a dataset of patients with SCD or thalassemia who met the criteria set up by the workgroup.

Resources

Each institute brought its own unique qualities to this partnership. The PA DOH provided the newborn screening database to the partnership and the staff to assist in the coordination of the development of the linked database. The hospitals each brought a wealth of data about its patients with SCD or thalassemia who currently were being treated or had been treated in the past. CMU brought in the computer resource and de-identification technologies, database management, and data linkage expertise to the partnership. Coordinating the process of creating the database required frequent conference calls with the partners, and time and effort dedicated to the project beyond that spent tracking patients and completing usual duties.

CMU's Management Science & Healthcare Informatics research group was instrumental in supporting the project through its expertise in data privacy, cleaning, and linkage. It was absolutely necessary to have the group's expertise to harmonize all of the data sources. Because all of the data already existed within the three childrens hospitals and the PA DOH, there were no costs incurred to obtain the data.

Lessons Learned:

Working with partners to establish timelines for data submission can be a very challenging process, especially when integrating numerous sources of data from different software platforms and dealing with different institutional policies and practices. Allowing for plenty of time and flexibility to discuss and resolve any problems that arise in the process is essential. Assembling the appropriate teams with expertise in the clinical domains; epidemiology and public health; information technology as applied to health care data and resources; analytic capabilities and project management; and, most importantly, knowledge of and links with the target community of patients with SCD also is essential to the success of such projects. One of Pennsylvania's limitations regarding this database was that data that were related to the adult population who had SCD or thalassemia were not included. Future collaborations will be able to obtain data related to adults with these diseases.

Outcomes

The formation of a partnership that was integral in accomplishing the construction of a hemoglobin database containing data from three separate treatment facilities and PA DOH's newborn screening database was a tremendous accomplishment for the state. Participating in this process provided a close to accurate account of the population of people with SCD or thalassemia living in Pennsylvania, especially the pediatric population, and the service use of treatment centers and community-based organization resources that are funded through contract agreements with the state. A stronger foundation to assess resource needs for the populations with SCD and thalassemia also was established.

From the International Classification of Disease, Revision 9 codes included in the database from each hospital, the PA RuSH team was able to translate these codes into the types of hospital services that were used by the pediatric population with SCD. This information then can be translated into actual dollars that can be used in a cost analysis (i.e., actual cost of average admission vs. insurance reimbursement; average cost of fever admission vs. reimbursement).

The PA DOH now has a more accurate snap shot of the populations with SCD or thalassemia in Pennsylvania. There also is a better understanding of the data linkage methods and mechanisms needed to aggregate statewide data about service use and resources needed for specific diagnosis groups. As a result of the work of the partnership, it will be possible to create a data hierarchy that can be used as a tool to determine what information or data are most helpful in understanding specific population needs.

As grantees of the PA DOH Sickle Cell Service grant, the CHOP, CHIP, and sickle cell hemoglobinopathy centers (SCHC) are required to provide quarterly reports to demonstrate sickle cell management and psychosocial service use. Through participation in RuSH, it was recognized that the existing reporting template could be modified to provide a more comprehensive snapshot of the patient population with SCD, and its service and resource use in the state of Pennsylvania. More specifically, the process of compiling surveillance data presented an opportunity for the PA DOH and its grantees to revisit current quarterly reporting fields and assess whether the right questions were being asked to determine service and resource use and create a data hierarchy as a tool to address specific circumstances (e.g., addressing populations needs or allocating funds). The data collection process also has led to the increase of statewide understanding of the populations with SCD or thalassemia and the associated service use data.

In addition, the process has emphasized the need to store newborn screening information and to maintain hospital system disease-specific databases until more innovative information technology systems are available. This also will help guide or modify the information collected for quarterly state reporting and answer questions such as: Are the appropriate data being provided or do modifications need to be made?

Pennsylvania also is well positioned to conceptualize and develop a statewide registry for SCD and thalassemia that can build on the integrated solution developed for the current effort. RuSH objectives that were accomplished from this data linkage project included collaboration, by involving appropriate within-state partners, and data integration, by linking information from the three childrens hospitals, the PA DOH's newborn screening data, and the state's vital records.

Sustainability/Translation

While the PA RuSH team has been successful in obtaining data from three hospitals catering to the needs of a large number of children with hemoglobinopathies, gaps in surveillance remain. Ongoing efforts to secure data user agreements between the DOH, PHC4, and Department of Public Welfare (DPW) will enable these data to be linked. Meetings with the DPW concerning these issues currently are under way. Discussions with the PHC4 will take place in the near future.

The establishment of these partnerships hopefully will serve as a model to expand the surveillance program to include other hospitals in Pennsylvania, as well as information on the adult population with SCD or thalassemia. Successful integration of data from the PHC4 and the DPW would enhance greatly efforts to accomplish this model surveillance program goal.

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Florida: Creating a Physician Reporting Website

Program Overview

Members of the Florida RuSH team, designed a website that allows physicians throughout the state to submit laboratory confirmed sickle cell disease (SCD) and thalassemia patient information, including personal identifiers such as name, social security number, date of birth, home address, place of birth, sex, race, and ethnicity for accurate linking of multiple source datasets used in the RuSH project (hospital discharge data, vital statistics, Children's Medical Services, Memorial Healthcare System, and Medicaid) and to aid in case confirmation. The website is completely secure and entered data is accessible only by Florida Department of Health staff. A physician creates a profile and is assigned a site identification number. Physician support staff can create a subaccount linked to the physician for purposes of entering data on the physician's behalf. To ensure the accuracy of data input by staff, the physician logs into the website to review and confirm data that then are transmitted to Florida Department of Health staff.

Clinicians with the highest volume of patients were identified through the hospital databases and subsequently contacted by our team, who encouraged them to enter SCD and thalassemia patient data into the website. Physicians identified through churches and social groups were also invited to enter data. In addition, the Cooley's Anemia Foundation was contacted, and responded with a list of physicians in Florida who provide care to individuals with thalassemia.

Resources Needed

The creation of a physician reporting website required the support of an experienced Web designer. Discussions with staff knowledgeable in health informatics, record linkage, and epidemiology, as well as with physicians, also were needed to define the types and formats of the data to be collected. Using the recommendations provided during these discussions, the website was designed and built. Furthermore, a list of physicians who provided confirmatory testing for the state newborn screening program and clinicians with high volumes of patients with SCD or thalassemia was essential to target the appropriate physicians to provide data. Financial incentives of $25 per patient record entered were offered to each physician to offset the time and effort required on their part.

Lessons Learned

The biggest hurdles we overcame in the creation and implementation of a physician reporting website were understanding the challenges of a physician's busy practice and identifying dedicated staff to enter data. Data were entered when staff had an opportunity to do so. It was essential to identify the person in the physician's office who was responsible for this task and work directly with him or her to facilitate the process or suggest to the physician bringing in an outside person for the project.

Outcome

The physician reporting website was launched in February 2012. Technical support was provided to sites and the first data entry began in April 2012, with 120 inputs in the first 48 hours. A total of 159 cases have been reported since then. The highest volume (53.21%) of cases was represented by hemoglobin SS (HbSS) . There were 6 cases of thalassemia and 12 cases reported as "Other Variant" as the diagnosis, including persistent fetal hemoglobin, Diamond Blackfan anemia, HbSN Baltimore, and HbCC.

Sustainability/Translation

Results of specific physician-reported cases can be shared with the reporting physician and aggregate data can be shared with all physicians caring for this patient population to observe the course of a disease, to understand variations in treatment and outcomes, to examine factors that influence prognosis and quality of life, and to describe care patterns, including appropriateness of care and disparities in the delivery of care.

It is unlikely that the Florida RuSH project activities will be sustained without continued extra-mural funding. The budget situation for state government in Florida continues to require scaling back of vital programs and services, and very few new programs have been funded in the past three years. While we plan to utilize the results of the RuSH project for health education activities and will continue to work with providers as well as support groups, without funding for data acquisition,

linkage and integration, and analysis, the surveillance portion of RuSH will likely not continue unless a new funding mechanism is obtained.

Moving forward, the approach utilized by the Florida RuSH team has potential to extend to a number of other genetic disorders and health conditions that pose continual challenges to those affected across their lifespan. Our model that included public health, epidemiology and informatics, clinician leaders, and outreach seems well suited to tracking other conditions and could be useful for other rare disorders and blood-related diseases.

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Michigan: Creating a Statewide Hemoglobinopathy Long-Term Follow-Up Module

Program Overview

Since 1987, children identified with sickle cell disease (SCD) through Michigan's Newborn Screening Program have been followed from birth through 5 years of age by the State of Michigan. To provide more comprehensive follow-up through the lifespan, Michigan recently developed a Web-based hemoglobinopathy follow-up module in the already established Michigan Care Improvement Registry (MCIR).

The hemoglobinopathy MCIR application allows for improved programmatic communication and invaluable information for hemoglobinopathy surveillance. The primary objective was to develop a Web-based follow-up module to monitor:

8. Diagnosis of all newborns referred with positive screening results for SCD.
9. Initiation of penicillin prophylaxis.
10. Hemoglobinopathy education and social work services.
11. Self-reported complications, screenings, and treatments related to SCD.

Resources Needed

Michigan birth records have been linked with newborn screening records since 2007. This linkage is completed weekly in the Division of Genomics, Perinatal Health, and Chronic Disease Epidemiology at the Michigan Department of Community Health using Link Plus software to identify potentially unscreened infants. Cases are matched using birth certificate identifiers and other unique information such as the infant's name, mother's name, and infant's date of birth. All linked records are uploaded weekly into the MCIR and profiles for children with abnormal hemoglobinopathy screening results are created automatically in the MCIR hemoglobinopathy module with newborn screening results determining the default case diagnosis.

Computer programmers with a background in the immunization components of the MCIR were tasked with the design and creation of a customized MCIR module for hemoglobinopathies. Follow-up duties (e.g., tracking status of penicillin prophylaxis and documenting home visits and telephone calls) were integrated into the application based on already established follow-up protocols in collaboration with the Sickle Cell Disease Association of America, Michigan Chapter (SCDAA-MI). In Michigan, the SCDAA-MI is responsible for provision of comprehensive services to all newborns with hemoglobinopathies detected by newborn screening. Data fields were established and approved by Michigan's Hemoglobinopathy Quality Improvement Committee, an active group of pediatric and adult hematologists practicing across the state. Data fields include: newborn screening laboratory results, confirmatory test results, penicillin prophylaxis start dates, ongoing status of prophylaxis through age five, and self-reported health status information, including demographic variables and barriers to care.

Lessons Learned:

Development and acceptability of a Web-based MCIR hemoglobinopathy module was successful as a result of significant time, staff, and funds expended specifically for this project. Replication of the module might be challenging for other institutions due to both the infrastructure and resources that are required for implementation. Building a similar module with fewer interactive and automated mechanisms still could serve as an effective surveillance tool without full-scale integration of every follow-up, communication, and reporting activity. In Michigan, this Web-based registry proved extremely useful for communication across different communities and allowed for health results and concerns to be identified in real time. This Web-based component also will be essential for any future integration of this module in sickle cell clinics by hematologists.

Outcome:

Regional patient advocates working for the SCDAA-MI have been documenting all follow-up activities for their clients in the hemoglobinopathy MCIR module since January 2011. Case profiles also are being created continually for older clients with SCD, with the goal of having baseline information for all patients with SCD from birth through 25 years of age in the MCIR by 2013. Data from the module will be included in a comprehensive surveillance and data report on SCD in Michigan, future educational campaigns, and potential advocacy initiatives. Michigan will continue to validate the data through a link between health status assessment findings and Medicaid claims. The health status assessment findings will provide valuable feedback to Michigan's hematologists, serve as the basis for adapting clinical protocols, and stimulate future research and program initiatives in Michigan's hemoglobinopathy quality improvement committee.

Sustainability/Translation:

A transition will take place during the final 6 months of RuSH funding, changing the title of this self-reported data collection initiative to "Sickle Cell Voices". Ongoing feedback will be used for quality improvement measures as questions are revised and adapted in the coming year. With enough momentum behind the hemoglobinopathy MCIR module, further completion of health status assessments and tracking of routine follow-up through the MCIR will continue past the end of the RuSH project. The Michigan Newborn Screening Program is in a secure position to fund troubleshooting and technical support to ensure that the application continues to be used to its fullest capacity. Newborn screening staff will monitor data collection for quality assurance, creating periodic briefs and reports. Applications for future data analysis will endure as surveillance information from the hemoglobinopathy MCIR module continues to grow with time.

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New York: Case Ascertainment of Individuals With Hemoglobinopathies Not Identified Through Newborn Screening

Program Overview:

As a result of newborn screening (NBS), all babies born in New York State (NYS) since 1975 have been screened for hemoglobinopathies. However, individuals born before screening began, in another country, in another state, or outside of a hospital setting are not in the NBS database. Therefore, such individuals must be sought out to provide the most accurate count of people living in NYS with these disorders, as well as available clinical information related to those individuals.

To identify such cases, letters were sent to providers at 31 pediatric hemoglobinopathy specialty care centers (HgbSCCs) informing them about the RuSH project and asking for them to participate in clinical data review of any patients seen and treated at their centers during the period 2004–2009. Those centers interested in participating in RuSH were sent copies of the project protocol, the executive summary, a list of data elements to be collected, and a copy of the NYS Department of Health's Institutional Review Board (IRB) approval of the project. Due to Health Insurance Portability and Accountability Act (HIPAA) privacy laws concerning individually identifiable health information, all RuSH project tasks concerning collection of personally identifiable information were subject to review and approval by each institution's IRB. Follow-up was conducted with each of the providers by telephone or email, or both, over the next several months to guide interested providers in submitting protocols to each of their IRBs prior to moving forward with data review.

In addition, a letter from the NY State Commissioner of Health was sought to give the RuSH principal investigators the authority to collect data on patients with hemoglobinopathy for the purpose of establishing a patient registry. As a result, an order was issued under Public Health Law 206 (1)(j) in January 2012 doing so. Both documents were distributed with the aim of facilitating IRB approval at individual HgbSCCs.

Resources Needed:

The goal of this project was to identify clinical case patients while minimizing the burden of data collection to center staff. Labor shortage was a common hurdle to overcome at centers when working on case identification and data extraction. To assist with these concerns, RuSH project staff worked with providers at each center to ensure an effective IRB submission. In addition, RuSH project staff helped draft patient consent forms, develop data entry tools needed for data collection, and offered their own services to perform site visits to centers to assist with review of patient records. As such, the resources required to complete clinical data collection included a substantial time commitment from and collaboration among team members, travel expenses associated with site visits to centers, and software to design and build a user-friendly database with an intuitive form of interface for data entry of patient information.

Lessons Learned:

Outreach to providers and corresponding clinical data collection required a substantial time commitment, constant communication, and organization. Regular communication with providers and center staff throughout the IRB submission process and the subsequent data collection efforts were essential to ensure timely progress. It was essential to have all key documents (project protocol, letters, approvals, etc.) on file and accessible to external partners working with RuSH team members, as these documents provided the backbone of IRB submissions at individual centers. IRB approval of the RuSH project varied considerably among institutions. Therefore, it was also essential to be flexible and available when working with providers to assist them in preparing the materials needed for their IRB submissions, to assist in drafting documents as requested, and to provide answers to questions posed by IRB panelists.

It sometimes took months for an individual center to move through the steps from the initial decision to participate in RuSH, to an effective IRB submission, to performing case identification of patients. This preceded the actual data collection itself, which might have involved a site visit for record review and data extraction. It was important to allot enough time for this process. For future projects involving clinical data, it is recommended that—in addition to structuring the research design in compliance with privacy and HIPAA laws–a legislative mandate or letters from the proper authority granting access to clinical data be obtained. Also, additional labor and funding would be useful for the work involved in chart review and data extraction.

Outcome:

A summary of our outreach efforts follows: 31 pediatric centers were contacted initially regarding RuSH clinical data collection; 7 centers received IRB approval and have begun the case identification and data collection process; 9 other centers have shown interest in participation, but are at various stages in the IRB discussion and approval process; seven centers opted out of participation due to resource issues, such as time and labor; and no response was received from 8 centers after repeated attempts at contact. At each of the approved centers, the New York RuSH team was able to identify cases not previously identified through NBS. The clinical data, representing several hundred cases, should shed some light on the prevalence of hemoglobinopathies and the most common treatments administered to patients. The current list of IRB approved centers comprises: New York-Presbyterian/Weill Cornell Medical Center, New York Methodist Hospital, Brookdale University Hospital and Medical Center, Staten Island University Hospital, Interfaith Medical Center, Women & Children's Hospital of Buffalo, and Bronx-Lebanon Hospital Center.

Sustainability/Translation:

Now that the groundwork has been laid and contacts have been established with providers in treatment centers around NYS, and particularly in the New York City area, continued communication and collaboration might continue past the end of the RuSH project period. The next step following this work will be to reach out to adult HgbSCCs. In previous RuSH community outreach projects, the NY RuSH team administered surveys to providers at adult HgbSCCs to assess the long-term health care status of patients, determine what clinical information was collected by the centers, and explore the possibility of data collection and sharing of patient data with the RuSH project team. Based on the responses received, available clinical data from patients seen at these centers will be sought, following the same protocol established for outreach to pediatric centers. In future data collection efforts, funding would be an essential component to success, as staffing is necessary for the work involved in data review and data entry of hundreds of cases. Hospitals have limited resources to allocate to these tasks without funding for necessary staff.

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Gratitude is extended to the dedicated clinicians, health care providers, and community-based organizations that support individuals and countless families affected by hemoglobinopathies. Finally, every sickle cell disease and thalassemia client is appreciated for his or her participation in research and surveillance efforts that will change the course of health care and health policy for future generations

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Bronx-Lebanon Hospital Center
www.bronx-leb.org/

Brookdale University Hospital and Medical Center
www.brookdale.edu/

California Department of Health Care Services
Data and Research Committee

Management Information Services
www.dhcs.ca.gov

California Department of Public Health Genetic Disease Screening Program
www.cdph.ca.gov/programs/GDSP/Pages/default.aspx

Carnegie Mellon University
www.cmu.edu

Carolinas Medical Center
www.carolinashealthcare.org

Children's Hospital of Philadelphia
www.chop.edu

Children's Hospital of Pittsburgh
www.chip.edu

Columbia University Medical Center
www.cumc.columbia.edu/

Community Health Interventions and Sickle Cell Agency
www.communityhealthinterventions.org

Comprehensive Sickle Cell Center of Georgia Health Sciences University
www.georgiahealth.edu/centers/sicklecell/

Cooley's Anemia Foundation
www.cooleysanemia.org

Duke University Comprehensive Sickle Cell Center
www.sicklecell.mc.duke.edu

East Carolina University Brody School of Medicine
www.ecu.edu/med

Georgia Comprehensive Sickle Cell Center at
Grady Health System
www.gradyhealth.org/clinic/70/

Georgia Health Policy Center
aysps.gsu.edu/ghpc/

Interfaith Medical Center
www.interfaithmedical.com/

Memorial Healthcare System
www.mhs.net; www.floridasickle.org

Michigan Department of Community Health Division of Genomics, Perinatal Health, & Chronic Disease Epidemiology
www.michigan.gov/dgphcde

Mission Hospital
www.missionhospitals.org

Newborn Screening for Metabolic and Sickle Cell Disorders Program of Georgia Department of Public Health
health.state.ga.us/programs/nsmscd/

New York Methodist Hospital
nym.org

New York Presbyterian
nyp.org

New York State Newborn Screening Program
www.wadsworth.org/newborn/index.html

North Carolina Sickle Cell Syndrome Program
www.ncsicklecellprogram.org

North Carolina Governor's Council on Sickle Cell Disease and Related Disorders, Piedmont Health Services and Sickle Cell Agency
www.piedmonthealthservices.org

Sickle Cell Community Advisory Council of Northern California

Sickle Cell Disease Association of America, Inc.
www.sicklecelldisease.org

Sickle Cell Disease Association of America, Inc. Eastern North Carolina Chapter
www.sicklecelleasternnc.org

Sickle Cell Disease Association of America, Michigan Chapter
www.scdaami.org

Sickle Cell Disease Association of Florida
scdaflorida.com/

The Sickle Cell Disease Foundation of California
www.scdfc.org/

Sickle Cell Disease Program of Children's Healthcare of Atlanta
www.choa.org/childrens-hospital-services/cancer-and-blood-disorders/programs/sickle-cell-disease

The Sickle Cell Foundation of Georgia, Inc.
www.sicklecellga.org/

St. Christopher's Hospital for Children
www.stchristophershospital.com

Staten Island University Hospital
www.siuh.edu/

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publichealth.med.miami.edu/

University of North Carolina Comprehensive Sickle Cell Program
medicine.med.unc.edu/centers/unc-comprehensive-sickle-cell-program-1

University of North Carolina at Greensboro
Center for Social, Community, and Health Research Evaluation
www.uncg.edu/rsh/cschre.html

University of South Florida
www.usf.edu

Wake Forest University Baptist Medical Center
www.wakehealth.edu

Women and Children's Hospital of Buffalo
www.wchob.org/

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