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Issue Brief

State Newborn Screening and Birth Defects Program Roles in Screening for Critical Congenital Heart Defects (CCHD)

The AMCHP Role

The Association of Maternal & Child Health Programs (AMCHP) is a national resource, partner and advocate for state public health leaders and others working to improve the health of women, children, youth and families, including those with special health care needs.

AMCHP supports state maternal and child health (MCH) programs and provides national leadership on issues affecting women and children*. We work with partners at the national, state and local levels to expand medical homes; provide and promote family-centered, community-based, coordinated care for children with special health care needs; and facilitate the development of community-based systems of services for children and their families.*

Introduction

Critical congenital heart defects (CCHD), also known as critical congenital heart disease, is one of the most common birth defects among infants in the United States. CCHDs account for 30 percent of infant deaths due to birth defects; approximately 7,200 infants are born with CCHD every year in the United States.[1] Infants born with a CCHD are at a significant risk for death or disability if their condition goes undiagnosed soon after birth. These infants have an increased chance of developing serious complications within the first few days or weeks of life and require emergency care.[2] Those infants whose condition is diagnosed often require surgery or catheter intervention in the first year of life.[3] Pulse oximetry is a non-invasive, low-cost test that has been identified by both public health professionals and clinicians as the preferred method of screening for CCHD and is an effective way to reduce the number of infants who are undiagnosed with CCHD.[4] In September 2011, HHS Secretary Kathleen Sebelius recommended that CCHD screening be added to the Recommended Uniform Screening Panel in the United States.[5]

The role of public health agencies in CCHD screening is multifaceted, complex and variable across the nation. Public health departments and programs may provide or give consultation about long-term and ongoing screening activities as well as follow-up assessment.[6] Public health birth defects surveillance programs can help increase the quality of screening: they conduct preliminary epidemiologic studies of CCHD that contribute to program evaluation and monitoring of outcomes, providing ongoing and timely feedback to programs. In addition, birth defects surveillance programs can provide continuously updated population-based data with regards to CCHD screening. Finally, evaluation of program effectiveness through long-term follow-up activities and quality improvement also should be considered.[7]

State newborn screening programs offer a comprehensive and coordinated system of education, screening, follow-up, diagnosis, evaluation, and management.[8] In addition, state Title V MCH programs support both newborn screening and birth defects programs by routinely conducting public education, training health-care providers, and supporting newborn screening programs and services for children with special health care needs.[9] This issue brief highlights effective practices and policies for states, specifically roles for state newborn screening and birth defects programs, to consider when designing and implementing comprehensive CCHD screening programs.

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ASSOCIATION OF MATERNAL & CHILD HEALTH PROGRAMS

Recommendations for State Newborn Screening and Birth Defects Programs

In February 2013, AMCHP hosted a meeting of state newborn screening and birth defects partners to discuss current and potential roles for newborn screening and birth defects programs in point-of-care CCHD newborn screening. Representatives from 12 states (CT, GA, IN, ME, MI, MN, NH, NJ, UT, VA, VT and WI), at various stages of CCHD program implementation, and partners identified effective practices and policies to implement and evaluate CCHD screening programs.

Key Elements of Successful Programs

States should take into account three key elements when moving forward in program development and implementation: **public health involvement** in the planning, implementation and follow-up processes; **data considerations** for collection and evaluation; and appropriate **stakeholder engagement** in the process.

Public Health Involvement

Newborn screening and follow-up is a public health role, and public health provides leadership for interagency or interorganizational partnerships that are needed to implement comprehensive, effective programs. Effective screening and follow-up services for CCHD requires partnerships between hospitals, clinical and community agencies and organizations, and state public health departments. Public health and newborn screening programs may have the capacity and infrastructure to lead or assist in the development and implementation of CCHD screening programs inclusive of education, screening, short- and long-term follow-up, diagnosis, systems to support hospital-based data collection, management, evaluation and quality assurance. Public health programs often have existing relationships with key partners, such as hospitals and birth facilities, community coalitions, and parent organizations. In operational screening programs, public health agencies and Title V programs play a central role in the implementation of a well-designed program to screen for, ensure prompt care and connections to resources and long-term follow-up, and improve health outcomes for infants with CCHD and their families.

Furthermore, it is important to promote the principals of population-based screening to the hospitals and facilities that will be implementing the point-of-care screenings. Public health can take the lead in developing or adapting evidence-based protocols and standardized algorithms for screening in a state, as well as providing guidance around technologies, mechanisms for reporting, developing implementation tools and communication resources for families. Several states have found success in establishing recommended protocols and providing training to hospitals and birthing facilities.

Newborn Screening and Title V

Newborn screening is a public health success story, enabling the early identification and treatment of infants with potentially life-threatening heritable disorders and genetic diseases. In the United States, roughly four million infants are screened each year. In 2002, some states were screening for only four conditions, while others were screening for up to 36. Today, 44 states and the District of Columbia require screening for at least 29 of the 31 core conditions on the Recommended Uniform Screening Panel. This widespread expansion and standardization of newborn screening has led to treatments and interventions for at least 12,500 newborns diagnosed with genetic and endocrine conditions, and hearing loss each year.

The Title V MCH Services Block Grant authorized by the Social Security Act is the only federal program of its kind devoted solely to improving the health of all women and children. The Title V MCH Block Grant is critical to state efforts in implementing systems of care for children with special health care needs. Historically, funding from this program supported newborn screening, diagnosis, treatment and follow-up care. State children and youth with special health care needs (CYSCHN) directors are often members of state newborn screening advisory committees. State Title V programs also have a long history of facilitating care coordination to vulnerable populations. Compared with other state services, Title V programs often have the greatest experience reaching the CYSHCN population, a strong connection of networks of pediatric specialists and the best data on the service needs of CYHSCN and their families.

- **Legislative considerations –** Many state legislatures approved legislation or are considering legislation requiring all birthing hospitals to screen for CCHD. While many hospitals voluntarily initiate routine screening for CCHD, the legislative mandate provides a foundation and driver for comprehensive CCHD screening programs. If state legislatures pursue mandating screening for CCHD it is recommended that data collection and reporting with supporting resources be part of the mandate. Some states have also found it helpful to specifically include a leadership role for health departments to collect data in the legislation to help establish quality improvement measures and surveillance efforts. This allows for the opportunity to put uniform protocols and systems in place to track data on health outcomes and evaluate the efficacy of services. Prior to mandating screening for CCHD, it is important for each state to consider the current readiness of hospitals and public health systems to ensure programs have the capability, or enough time and resources to develop the capability, to comply with the mandate.

  For states that use the authority of health department advisory committees or the regulatory process to add CCHD or other new conditions to newborn screening panels, it is recommended the same data collection, reporting and quality improvement measures be included.

Data Considerations

Quality data serve as the foundation to successful programmatic implementation and evaluation. Most newborn screening programs currently screen the majority of conditions through a heel stick test, with blood spot analysis occurring at public health or contracted laboratories. Pulse oximetry screening for CCHD is a point-of-care test that occurs in hospitals before an infant is discharged. Point-of-care tests can present challenges in data collection for appropriate public health surveillance as uniform reporting systems may not be established with hospitals and birthing facilities. Data collection provides an important area for collaboration between programs, particularly newborn screening and birth defects.

CCHD screening results are entered into the hospital medical record. Birth defects surveillance programs often draw from hospital medical records, and can provide this data to newborn screening programs for retrospective analysis. In addition to data sharing, there are other opportunities for collaboration and development of a data system, such as tracking and evaluating screening outcomes. Most birth defects surveillance programs already collect data to calculate CCHD prevalence, and also can assist in quality improvement by using their current data collection systems to evaluate false-positive and false negative screens for CCHD.[10] Monitoring and evaluating- short and long-term follow-up also is an important aspect of data collection. In some states, Title V CYSHCN programs have systems in place to assess management for conditions; access to medical, community and family support services; and facilitate care coordination.

Lessons also are learned from the Early Hearing Detection and Intervention (EHDI) programs, which conduct hearing screening in the clinical setting. EHDI programs have identified the importance of collecting child-level data for quality improvement and have data systems already in place to collect aggregate data. These programs can help identify commonalities to work together with newborn screening for CCHD. Other key lessons learned from EHDI include the importance of integration with newborn screening programs and that state health departments should play a leadership role in implementing electronic data systems for timely reporting.[11]

While there is variation among states in terms of resources and methodologies to collect data, operational screening programs underscore the importance of having clear and appropriate definitions of the types of data the programs want to collect when moving forward with implementation. National resources and assistance are available to help states form data collection systems for CCHD. NewSTEPs (Newborn Screening Technical assistance and Evaluation Program) is a comprehensive newborn screening and technical program designed to provide data, technical assistance, and training to newborn screening programs across the country and to assist states with quality improvement initiatives. NewSTEPs, a Health Resources and Services Administration (HRSA)-funded program at the Association of Public Health Laboratories, has worked with newborn screening program experts to develop national quality indicators, and to propose national CCHD data elements for states.

Similarly, the National Birth Defects Prevention Network (NBDPN) is a national organization of population-based birth defects programs and individuals interested in birth defects surveillance, research and prevention; NBDPN serves as a resource to improve the quality of birth defect surveillance data and provides technical assistance for the development of uniform methods of data collection. NBDPN has developed public health surveillance case definitions for CCHDs and guides state programs in monitoring and evaluation.

Stakeholder Engagement

Strong collaborations are essential to effectively implementing CCHD screening and follow-up programs. It is vital for state programs to engage key stakeholders in the beginning of the process. Newborn screening and birth defects surveillance programs have an important role collaborating with each other throughout the process. Additionally, state Title V children and youth with special health care programs can potentially play a role in facilitating care coordination, ensuring follow-up and collecting data to promote quality improvement. State health departments can also facilitate engagement and collaboration with EHDI and Vital Records as those programs may have data and systems in place to work together with state newborn screening (NBS) programs on point-of-care screening. Stakeholders can also include health care providers and consumer groups who have long worked with both newborn screening systems (blood spot and point-of-care) and birth defects surveillance programs.

Because CCHD is a point-of-care screening, strong collaboration with hospitals and birthing facilities are critical. Agencies such as state hospital associations can help foster connections and can be an important partner in the process. It also is important to connect with pediatric cardiologists and technicians who make the final diagnostic determination and provide these specialists training on the importance of data reporting, screening follow-up and long-term care. Establishing working agreements with hospitals and birthing facilities is a helpful strategy to ensure access to echocardiography data and follow-up for all newborns with possible CCHD.

Other key stakeholders are parents and policymakers. Health promotion activities provide education and messages to families and communities. Core messages for health promotion activities should include awareness of CCHD, reinforcement of the benefits of newborn screening and information on the treatment for the condition if identified. State health departments may provide early education and resources on the importance of screening for CCHD and having comprehensive screening and follow-up programs, depending on staffing and budgetary considerations. Parents are often powerful advocates for newborn screening and can work with health departments to help disseminate important information and resources. As legislators develop policies, it is important to understand the resources and factors involved in effective screening and follow-up programs to ensure legislation is supportive of comprehensive screening programs for CCHD.

State Case Studies

A map of newborn CCHD screening progress in the United States can be found at cchdscreeningmap.org/ and under the “Newborn Screening for Critical Congenital Heart Disease” heading at aap.org/stateadvocacy.

The following case studies represent four states that have established strong collaborations to successfully incorporate a CCHD screening program. Each case study highlights those collaborations and details the state approach to program implementation, key elements of the approach that contributed to a strong foundation, how data is collected, and program implementation challenges and successes.

INDIANA

Approach

In 2011, the Indiana General Assembly passed legislation to include CCHD screening as part of the state-mandated newborn screening panel. Effective January 2012, all birthing facilities in the state were required to perform pulse oximetry screening on all newborns to detect critical congenital heart defects. Prior to implementation, the Indiana State Department of Health (ISDH) Genomics & Newborn Screening Program, which is housed under the Title V authority, began working with neonatologists, nurses, pediatric cardiologists, and high-risk obstetricians to finalize the screening protocols; identify guidelines and recommendations

related to purchasing, upgrading, or standardizing pediatric pulse oximetry equipment; and identify the type of data that would be required for reporting to ISDH. To help assess the ability of the state to implement CCHD screening, ISDH sent surveys to birthing facilities.

A screening protocol was determined and disseminated to primary care physicians, midwives, OB/Labor & Delivery/Nursery department managers at each birthing facility, genetic counselors, geneticists, pediatric cardiologists, and the newborn screening coordinator at each birthing facility. A CCHD fact sheet was developed and information, including the Indiana CCHD protocols, was posted to the ISDH website. ISDH also developed parent education materials, which were reviewed by the parent of a child with a heart defect detectable by CCHD screening.

Key Elements

- Legislative mandate
- Early Involvement of public health in the program design
- Involved multiple stakeholders in clinical practice and parents from the community
- Established recommended protocols
- Developed parent/community education

Collaborations

The ISDH worked with hospitals and birthing facilities to implement the program and retrieve screening data from all birthing facilities. ISDH expanded the Indiana Newborn Screening Tracking & Education Program (INSTEP) Web-based application, developed in-house, which allowed for the facilitation of tracking activities by ISDH. This application allows birthing facilities to report to the newborn screening program children who did not receive a screen or children who did not pass a screen. ISDH also collaborated with Indiana Perinatal Network to distribute all CCHD-related messages to OB/L&D/Nursery managers.

As noted in the next section, the newborn screening and birth defects programs collaborate on data collection in order to ensure the receipt of follow-up information, including health outcomes, on children who did not pass a valid heel stick or CCHD screen.

Data Collection

The INSTEP Web-based application currently in use by all birthing facilities was used to submit monthly reports regarding the heel stick screen. The ISDH newborn screening program expanded the required monthly summary reports to include CCHD screening. The reports include detailed information from birth facilities on infants born in Indiana who did not pass the CCHD screen or did not receive a valid CCHD screen.

To obtain follow-up information, including health outcomes, on children who did not pass CCHD screening, INSTEP has partnered with Indiana Birth Defects & Problems Registry (IBDPR), which is another Web-based application developed, maintained, and used by the Genomics & Newborn Screening Program. IBDPR collects information on birth defects for all children born in Indiana from birth up to age three. Data is obtained from direct physician reporting, hospital discharge data and medical record audits. IBDPR data also is used to ensure that all children who did not pass the CCHD screen received timely and appropriate follow-up care, and to evaluate health-related outcomes for children who are diagnosed with at least one of the critical congenital heart defects detectable by CCHD screening. Indiana also plans to use data to evaluate and potentially modify current standards of care for Indiana children with CCHD.

Challenges

- Lack of CCHD protocols for neonatal intensive care unit (NICU) babies
- Need for further education regarding current CCHD protocol and appropriate monthly reporting of babies that did not receive screens or did not pass screens
- There is a difference in newborn screening and birth defects registry laws. While all infants born in the state are screened; the birth defects legislation covers state residents, so follow-up and getting confirmatory information can be a challenge if the individual is screened or gets care in another state
- Refinement of plan for follow-up of newborns that have failed CCHD screen
- Need for further education in the midwife and homebirth community

Successes

ISDH has implemented a state-wide CCHD screening program so that all infants in the state are currently screened. As of Jan. 30, 2013, preliminary data indicated that Indiana had approximately 81,200 live births and of those 45 newborns did not pass the pulse ox screen. Of the newborns that did not pass, 25 received an echo at a birthing center, 17 received an echo at an in-state

hospital, and three received an echo at an out-of-state hospital. According to the preliminary data, from the 45 newborns that did not pass the pulse ox screen, 12 were reported to the IBDPR with any heart defect and five were reported to the IBDPR with one of the seven targeted heart defects.

The current ISDH integrated data system was expanded to allow ISDH to receive monthly data from hospitals regarding CCHD screening and tracking information. Collaborations within department programs provide Indiana with the opportunity to track follow-up and outcomes for those infants with a CCHD. Evaluation of the best available methods for collecting data is currently underway.

MICHIGAN

Approach

At the present time, pulse oximetry screening for CCHD is not mandated in Michigan; however, the Newborn Screening Advisory Committee recently recommended adding it to the screening panel beginning in 2014. Currently, screening is done on a hospital-by-hospital basis and most hospitals have already begun to include pulse oximetry screening as standard of care for all well-babies. The Michigan Department of Community Health (MDCH) is implementing a HRSA-funded demonstration program to assess feasibility and implement statewide pulse oximetry screening of all newborns for critical congenital heart defects prior to hospital discharge. The goals of the demonstration project are to increase the number of Michigan newborns screened for CCHD using a validated screening protocol, and to develop state infrastructure for collection of CCHD screening data through electronic health information exchange to enable effective public health follow-up, quality assurance and evaluation.[12] A growing number of hospitals have volunteered to use a standard screening algorithm and participate in a pilot project for collection and reporting of pulse oximetry data.

In 2012, the MDCH established a CCHD Advisory Committee to provide clinical guidance and expertise in development of a screening algorithm, identification of key data elements, and follow-up protocols. The committee also helps to identify ways of addressing barriers to screening and early treatment. Members include pediatric cardiology sub-specialists, pediatric hospitalists, neonatologists, newborn nursery nurses, advocacy organizations and parent representatives from

across Michigan. This committee will remain a critical resource for informed decision making for clinical protocol and reporting methods. To complement existing newborn screening and birth defects staff available to assist with implementation of CCHD screening, MDCH also hired a former NICU nurse to serve as a CCHD nurse educator providing training and technical assistance to hospitals.

Key Elements

- Early Involvement of public health in the program design
- Using a team approach that enhances collaboration between newborn screening and birth defects programs
- Establishing recommended protocols and reporting methods specific to Michigan
- Involving multiple stakeholders from hospitals, cardiology, advocacy group and family representation
- Developing a public health database for tracking CCHD results that interfaces directly with the existing NBS system for identification of cases needing follow-up

Collaborations

The Michigan pulse oximetry newborn screening program is administered by the MDCH, Genomics and Genetic Disorders Section, which has responsibility for both the blood spot screening and birth defects follow-up programs. Therefore, both newborn screening staff, as well as the birth defects program coordinator, are actively involved and working together on all aspects of the program ranging from development and dissemination of CCHD information and education materials to data system infrastructure. Through the CCHD Advisory Committee mentioned above, as well as the internal Birth Defects Steering Committee, Michigan has successfully tied the birth defects registry and Title V CYSHCN program in with

development and long-term evaluation of the new screening program. In addition to using birth defects registry data and death certificates for identifying missed cases and assessing co-morbidities and long-term outcomes of children with CCHD, MDCH plans to collaborate with the three pediatric cardiac surgery centers in the state to obtain confirmatory diagnostic information related to heart surgeries and catheterizations.

The Michigan CCHD newborn screening program also initiated contact with a majority of birthing hospitals through previously identified NBS coordinators, as well as new contact persons identified through a statewide survey, on current status of CCHD screening. Direct feedback from hospitals as well as through the advisory committee has provided valuable information on barriers and solutions to implementation of pulse oximetry screening.

Data Collection

The CCHD screening program was classified as "not human subjects research" by the MDCH Institutional Review Board (IRB). Accordingly, several hospitals willing to submit screening data received rapid approval from their respective IRBs allowing submission of screening results to MDCH. After evaluating various options, MDCH decided to develop an electronic reporting module for reporting of all pulse oximetry screening and CCHD-related follow-up taking place in hospitals. Hospitals will report patient-level pulse oximetry results for all newborns according to the Michigan algorithm based on national Centers for Disease Control and Prevention (CDC) and American Academy of Pediatrics (AAP) recommendations. Echocardiogram and case follow-up information also will be documented along with surgical procedures. Hospital data submission to MDCH will begin on a rolling basis as hospitals complete modifications to their electronic medical records (EMRs) to enable collection of pertinent data variables, and for the larger institutions, as EMR reports are developed that can be uploaded to a secure FTP site rather than entering patient data individually. Long-term plans include development of capabilities for direct data exchange using HL7 messaging through the state Health Information Network.

To identify children that are missed by screening, Michigan plans to link pulse oximetry screening results with blood spot screens and birth certificates to identify infants who were not screened and provide feedback to birthing hospitals to assist with quality assurance. The sensitivity, specificity and positive predictive value of the screening algorithm also will be evaluated to identify the need for any changes to the screening protocol.

Challenges

- Concerns about extra staff time for reporting results to MDCH, even among hospitals already screening
- Lack of state mandate for screening; hospitals report it will be easier to justify reporting/ allocation of staff time once universal screening required
- Need for hospitals to purchase new/additional screening equipment
- Need to follow screening procedures consistent with state recommended algorithm – has resulted in some hospitals changing current practices
- Need to clarify screening procedures for NICU babies
- Delays in achieving changes to state IT systems and servers required for implementation of data collection using the new pulse oximetry screening module
- Need to identify referral patterns for pediatric echocardiogram for infants with a positive screen born in hospitals with limited access to pediatric cardiology

Successes

Michigan experienced many successes with implementation of CCHD screening despite not yet being mandated in the state. Overall, birthing hospitals have been enthusiastic and receptive to screening, with several modifying their procedures to comply with the state algorithm. There was a good response to a statewide survey on current pulse oximetry screening practices, which showed that more than 80 percent of hospitals are currently screening, and all but a few plan to begin screening in the near future. Grant funding was used in the spring of 2013 to award 19 mini-grants to hospitals for implementation of pulse oximetry screening. The mini-grants were well received by hospitals and used in a variety of ways depending on the particular needs of a hospital, and included purchase of pulse oximetry equipment and/or supplies, staff training on screening techniques and proper data entry, parent education materials, and EMR modifications necessary for data collection and submission to MDCH. Finally, a new pulse oximetry data module has been developed, and plans are in place for using birth defects registry and other data sources to monitor screening at the

state level and assess outcomes of children identified by screening.

NEW JERSEY

Approach

In 2011, following a law signed by Gov. Chris Christie, New Jersey became the first state to implement newborn screening for critical congenital heart defects. Staff from the Special Child Health and Early Intervention Services (SCHEIS) – the Title V CYSCHN Program – along with CDC, HRSA, AAP – New Jersey Chapter, and the New Jersey Critical Congenital Heart Defects Screening Working Group worked to support and guide implementation efforts to build an effective CCHD screening program by partnering with birth defects surveillance. Implementation efforts have included identifying contacts at every birthing facility in the state, distributing a recommended screening protocol to the birthing facilities, providing trainings, and developing a parent education handout. Additionally, a surveillance mechanism was established and implemented to evaluate the impact of statewide pulse oximetry legislation.

Key Elements

- Legislative mandate
- Early Involvement of public health in the program design
- Existing collaborations between public health (Newborn Screening and Genetic Services Program/Birth Defects Registry) and hospitals
- Establishing recommended protocols
- Provider trainings and parent education

Collaborations

From the beginning, collaborations were critical. Networks already existed between New Jersey Department of Health (NJDOH) and the hospitals because of the Birth Defects Registry (BDR) and the Newborn Screening and Genetic Services Program (NBSGS). Nevertheless, prior to implementation, NJDOH requested that each hospital designate a “pulse ox liaison” to serve as a point of contact. NJDOH staff are in touch with these individuals on a formal and informal basis (e.g., letters, calls, webinars).

Further, the NBSGS and BDR Program both sit in the Special Child Health and Early Intervention Services Unit. While the medical director of Newborn Screening and Genetic Services oversaw the implementation of statewide CCHD screening, the two programs have worked quite closely on this initiative. The Birth Defects Registry Program manager and a birth defects registry nurse have both been active members of the Department of Health CCHD Core Team and essential collaborators on the HRSA CCHD Demonstration Grant. The BDR team has a longstanding relationship with the hospitals and this helped to facilitate implementation and data reporting. All abnormal screens, along with relevant data, are reported to the BDR.

Data Collection

New Jersey collects quarterly aggregate data from the hospitals (e.g., number born in the quarter, number screened and any discrepancies), and uses the BDR to collect data on all positive screens. Aggregate data are collected on all infants and individual level data are collected on screen positive infants. The NJ Electronic Birth Certificate (EBC) is in the process of being reengineered. Once completed, it is anticipated that NJ will collect individual-level data on all infants.

Challenges

- While statewide collaboration has been immense, limited initial resources presented challenges with implementing all aspects of a comprehensive screening program. In addition, the statewide data collection system took time to establish.

Successes

In the first nine months following implementation of the mandate to screen newborns for CCHDs using pulse oximetry, approximately 99 percent of the 73,320 eligible infants were screened for critical congenital heart defects. Through screening, hospitals detected three newborns with CCHDs. Without screening, the heart defects of these three infants might not have been found soon after birth, possibly resulting in death or disability.[13]

In terms of collaboration, the above programs collaborate extremely well together, and have always worked closely as they sit in the same unit. Having these systems and relationships already in place, as well as having previously established relationships between the NJDOH and the birthing facilities, helped to facilitate implementation of the new statewide CCHD screening program.

UTAH

Approach

Utah has 51 birthing facilities, of which 16 reported doing some form of screening for CCHD. Further follow-up indicates that only four Utah hospitals are doing any routine CCHD screening. One-fourth of birthing facilities are located in rural Utah, with 23 percent of births occurring in rural/frontier Utah. Utah received a three-year HRSA CCHD demonstration grant to pilot CCHD screening in two birthing facilities and to develop a statewide implementation plan. The Utah Department of Health (UDOH) has established coordinated efforts and assembled a core work group which meets monthly. The core group has agreed on using the Kemper screening protocol, what variables are to be collected as well as the education and training materials that are being used. An advisory committee of major stakeholders meets quarterly; members include the University of Utah Division of Pediatric Cardiology and General Pediatrics, Utah Hospital Association, Utah Chapter of the American Heart Association, Community Pediatricians, and bureau director of Children with Special Health Care Needs. As part of the grant, hospitals also will work with UDOH to conduct a cost analysis of screening implementation in newborn nurseries.

Recently, legislation was passed to mandate statewide screening for CCHD, which will go into effect October 2014. During the development of the legislation, UDOH staff worked with policymakers to include language that requires state pilots to determine high altitude protocols.

Key Elements

- Legislative mandate
- Early involvement of public health in the program design
- Establishing recommended protocols and reporting methods
- Existing collaborations between public health, specifically CYSHCN, Newborn Screening Program and Utah Birth Defect Network (UBDN), and Utah birthing facilities

Collaborations

The UBDN is a program within the UDOH and has existing relationships with birthing facilities in the state. UBDN serves to prevent birth defects and secondary disabilities by monitoring occurrence, conducting research, and providing education and outreach. UDOH has partnered with two hospitals to conduct a CCHD screening project to determine pulse oximetry cut-offs for high altitudes as well as the feasibility of data transmission within Utah. The state newborn screening programs, along with UBDN, are located in the bureau of Children with Special Health Care Needs within UDOH. This organization has allowed for these programs to work closely in creating an efficient pulse oximetry screening model. Utah is planning to continue these internal and external partnerships as they move forward with the screening project.

Data Collection

Utah has created a functional reporting system between the pilot hospitals and UBDN for data collection that could be implemented statewide. Each hospital pilot site has finalized an internal database for onsite data entry.

Additionally, each pilot hospital is developing its own system for documenting costs, including increase in work force, equipment, and software development. There are plans to do a Time and Motion Study within each pilot site.

Challenges

- Utah is a high altitude state, therefore pulse oximetry measurements must be evaluated for consideration of the need to adjust the cut off values for CCHD screening and minimize both false positive and negative results.
- Utah has faced IRB challenges with regards to the implementation of CCHD screening pilots. UDOH IRB process took longer than anticipated and eventually, UDOH moved forward with a program implementation project which did not require IRB for data collection and analysis. Legislation which mandated these pilots allowed for this. The two pilot sites' IRB processes went smoothly.
- October 2014 is the date for mandated statewide CCHD screening to begin. This will require the plan for statewide implementation to be developed earlier than had been planned in the grant timeline.
- Currently, many of the Utah birthing facilities are in rural/frontier communities with no local access to newborn echo cardiology; if statewide implementation was mandated now, 78 percent of newborns with positive screens would need to be transported outside their surrounding community for echocardiograms. The pediatric cardiology division is aware of the need to expand access and is working diligently to address these issues.

Successes

Utah achieved many successes with regard to collaborations. Key players from UBDN and UDOH, the University of Utah General Pediatric and Pediatric Cardiology Divisions, and the largest private health care provider in the state are working closely together. All core workgroup members attend monthly project meetings and the quarterly advisory committee meetings. Organizations involved in the implementation of CCHD screening have been very willing to help with the project. Pilots have begun at the two planned sites and any internal implementation problems are being actively addressed. Preliminary downloading of screening data indicates that the goals for data collection and analysis will be reached within the time frame of this three year grant.

Conclusion

States are moving forward in legislative and program activity to implemented universal newborn screening for critical congenital heart disease. This presents an opportunity for collaboration between state newborn screening and birth defects programs. The findings in this document can be used by states to inform planning and implementation of comprehensive, effective CCHD screen and follow-up programs.

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Additional Resources & Publications

- **Secretary's Discretionary Advisory Committee on Heritable Disorders in Newborns and Children**
  hrsa.gov/advisorycommittees/mchbadvisory/heritabledisorders/
- **CDC: Screening for Critical Congenital Heart Defects**
  cdc.gov/ncbddd/pediatricgenetics/cchdscreening.html
- **Newborn Screening Technical Assistance and Evaluation Program (NewSTEPs)**
  newsteps.org
  National Birth Defects Prevention Network
  nbdpn.org
- **Association of State and Territorial Health Officials (ASTHO) – CCHD Fact Sheet**
  astho.org/critical-congenital-heart-disease-issue-brief/
- **Report: An evidence development process for newborn screening**
  hrsa.gov/advisorycommittees/mchbadvisory/heritabledisorders/reportsrecommendations/reports/evidencereviewprocess.pdf
- **CDC MMWR Feature from Georgia and New Jersey** (pgs. 288-294)
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