Critical Congenital Heart Disease Newborn Screening Implementation: Lessons Learned

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Abstract

Introduction—The purpose of this article is to present the collective experiences of six federally-funded critical congenital heart disease (CCHD) newborn screening implementation projects to assist federal and state policy makers and public health to implement CCHD screening.

Methods—A qualitative assessment and summary from six demonstration project grantees and other state representatives involved in the implementation of CCHD screening programs are presented in the following areas: legislation, provider and family education, screening algorithms and interpretation, data collection and quality improvement, telemedicine, home and rural births, and neonatal intensive care unit populations.

Results—The most common challenges to implementation include: lack of uniform legislative and statutory mandates for screening programs, lack of funding/resources, difficulty in screening algorithm interpretation, limited availability of pediatric echocardiography, and integrating data collection and reporting with existing newborn screening systems. Identified solutions include: programs should consider integrating third party insurers and other partners early in the legislative/ statutory process; development of visual tools and language modification to assist in the

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interpretation of algorithms, training programs for adult sonographers to perform neonatal echocardiography, building upon existing newborn screening systems, and using automated data transfer mechanisms.

Discussion—Continued and expanded surveillance, research, prevention and education efforts are needed to inform screening programs, with an aim to reduce morbidity, mortality and other adverse consequences for individuals and families affected by CCHD.

Keywords

Critical congenital heart disease; Newborn screening; Public health

Introduction

Congenital malformations are one of the leading causes of infant death in the United States (U.S.), and critical congenital heart disease (also referred to as *critical congenital heart defects* [CCHD]) is responsible for more deaths than any other type of malformation (Hoffman and Kaplan 2002; Reller et al. 2008). It is estimated that approximately two of every 1000 live births will have CCHD; these conditions require surgery or catheter intervention during the first year of life (Mahle et al. 2009). Some cases are detected by a prenatal ultrasound; others may be detected on the basis of physical examination findings (e.g., heart murmurs, rapid breathing, respiratory distress, or blue skin) in the newborn, yet not all infants with CCHD are identified prior to hospital discharge (4–31%) (Brown et al. 2006; Liberman et al. 2014; Peterson et al. 2014a), which may result in significant morbidity or mortality. Pulse oximetry has been implemented as a point-of-care newborn screening test for CCHD, to identify these undiagnosed cases (Mahle et al. 2009). Performed 24–48 h after birth, pulse oximetry screening is a non-invasive measurement of the proportion of hemoglobin in blood that is saturated with oxygen. The presence of low blood oxygen saturation (hypoxemia), or a difference between pre-ductal and post-ductal (proximal and distal to the aortic opening of the ductus arteriosus, respectively) saturation, frequently precedes other signs or symptoms in infants with unrecognized CCHD (Kemper et al. 2011).

The primary targets of CCHD screening are seven specific lesions: (1) hypoplastic left heart syndrome, (2) pulmonary atresia, (3) tetralogy of Fallot, (4) total anomalous pulmonary venous return, (5) transposition of the great arteries, (6) tricuspid atresia, and (7) truncus arteriosus (Kemper et al. 2011). An additional five cardiac malformations (coarctation of the aorta, Ebstein's anomaly of the tricuspid valve, interruption of the aortic arch, double outlet right ventricle, and other single ventricle heart disease) have been detected in pulse oximetry screening studies less consistently, and may be included by screening programs as secondary targets [Mahle et al. 2009]. The screening characteristics (e.g. detection rate, false negatives) vary by CCHD type.

As part of the Newborn Screening Saves Lives Act of 2008, the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) was established to reduce morbidity and mortality in newborns and children that have, or are at risk for, heritable disorders (Appendix 1) (Hoffman et al. 2002). Following an evidence review process, SACHDNC recommended to the Secretary of Health and Human Services to add

CCHD to the Recommended Uniform Screening Panel (RUSP) for NBS (Secretary's Advisory Committee on Heritable Disorders in Newborns and Children 2015); the Secretary adopted the recommendation in September 2011 (Sebelius 2011), which was endorsed by the American Academy of Pediatrics (Mahle et al. 2012). In her recommendation, the Secretary charged the Health Resources and Services Administration (HRSA) to develop screening standards and infrastructure needed for a public health approach for point-of-care CCHD screening.

In response to the Secretary's charge, HRSA funded six CCHD screening demonstration projects (funding ($300,000 per year) for a 3 year period, June 2012–May 2015). These projects were charged to support the development, dissemination and validation of screening protocols and infrastructure needs for point-of-care screening specific to CCHD. The six funded programs were: (1) Michigan; (2) New Jersey; (3) Utah; (4) Virginia; (5) Wisconsin; and (6) a consortium of five New England states (Maine, New Hampshire, Vermont, Rhode Island, and Connecticut). The purpose of this report is to present the collective experiences of these grantees, highlight issues that may be important to those interested in CCHD screening programs, and describe the successes and challenges to assist policy makers and public health programs that are considering, or are in early stages of, CCHD screening implementation.

Methods

In HRSA's Funding Opportunity Announcement (Catalog of Federal Domestic Assistance number 93.110), each of the grantees was required to: (1) create a network including the state public health department, hospitals and/or birth facilities; (2) develop a plan to incorporate CCHD screening and reporting at the provider and state levels; (3) develop guidelines to collect and report results of CCHD screening to stakeholders (e.g. state public health department, families); (4) develop and deliver educational programs about CCHD screening, counselling, testing, followup, treatment and specialty services to parents, families, and patient advocacy and support groups, (5) perform quality assurance, outcomes analyses, cost analysis, and other public health monitoring functions; (6) establish guidelines for follow-up methodology and reporting; (7) coordinate with Maternal and Child Health Bureau-funded programs; and (8) establish, operate, and maintain a state-level electronic system to systematically collect and analyze data, and evaluate the impact of CCHD screening (e.g., proportion of babies screened prior to discharge, number of CCHD cases detected by screening, health care utilization). In 2013, the Newborn Screening Technical assistance and Evaluation Program (NewSTEPs) began hosting a monthly technical assistance conference call for individuals interested in CCHD screening, and in February 2014, partnered with the Pediatric Congenital Heart Association to convene an in-person meeting to discuss the current status of CCHD screening in the U.S., and to provide ideas and guidance for screening programs across all stages of implementation. The grantees and other leaders in CCHD screening shared challenges and successes with invited representatives. Following the February meeting, continued education efforts include monthly webinars and several video conferencing sessions for all demonstration projects, hosted by Virginia's CCHD Screening Program. This report is a product of these collaborations.

Results

Legislation and Sustainability

While the SACHDNC recommends that all conditions listed in the RUSP be integrated in every newborn screening program, which conditions each state includes, as well as their process for adding conditions, varies; some require legislation, while others use a regulatory process. State requirements for implementation and the status of states that have adopted CCHD screening as of March 12, 2015 are available (Appendix 1) (Reller et al. 2008; Glidewell et al. 2015). Legislative and statutory mandates for screening, data collection and reporting processes and requirements for state newborn screening programs are not uniform. A number of state programs lack the public health authority to collect data on individual newborns, underscoring the importance of ensuring that data collection is included in the legislative language.

A common challenge of adding new disorders to screening programs is a lack of funding and other resources. Most screening programs use newborn screening fees to support broad program activities. Some stakeholders pursued a fee increase via legislation or other regulatory mechanisms to support CCHD screening implementation. In addition, some programs have explored non-traditional funding approaches such as tobacco taxes. Further, for both initial implementation challenges as well as long term sustainability, programs should consider integrating third party insurers and other partners early in the developmental process. Finally, adoption of a Current Procedural Terminology (CPT) code from the American Medical Association could also help to sustain CCHD screening by providing unique reimbursement.

Screening Algorithms and Interpretation

The recommended screening algorithm endorsed by the American Academy of Pediatrics (AAP) (Kemper et al. 2011) (Appendix 2) has been widely adopted. Some states (e.g., New Jersey) (Garg et al 2015) and birthing facilities have developed alternative algorithms to improve the outcomes of the screening algorithm locally. As screening implementation has occurred in many states, questions regarding the most appropriate cut-off values, need for both a pre-ductal and post-ductal reading, best timing of the screen, number of repeat screens, and requirements for failure of the screen have been raised. Studies, including mathematical modelling, are currently underway examining the impact of algorithm modifications on the screening characteristics.

The HRSA grantees commonly received feedback from birth registrars and nurses regarding the screening terminology and algorithm interpretation. This feedback resulted in projects implementing changes to modify the language to "negative/pass" and "positive/fail" to ensure more consistent reporting of CCHD screening results. In particular, recognition that greater than 3% differential between pre- and post-ductal results constitutes a failed screen has been documented as a common challenge. (Appendix 2). Education efforts and development of an enhanced visual tool based on the screening algorithm were strategies used to improve accuracy of interpretation of CCHD screening results and ease of use for healthcare providers (Fig. 1).

Children born at high altitude may have lower pulse oximetry levels than those born at sea level; thus there may be more false positives from CCHD screening at higher elevations. Grantees did not directly evaluate this issue. However, research is currently being conducted to determine if screening algorithms should be adjusted, based on elevation. There is evidence that alternative algorithms are warranted at moderate altitudes (2500–5500 feet) [Samuel et al. 2013; Wright et al. 2014).

Provider and Family Education

Education of both providers and parents is critical for successful implementation of CCHD screening. Comprehensive provider education includes information on benefits and limitations of screening, performing the screening, algorithm interpretation, and short- and long-term followup recommendations. Education for parents and family is necessary to increase awareness of CCHD screening using pulse oximetry and to ensure an understanding that not all CCHDs are detected with the screen (false negatives). Resources with appropriate literacy levels and in languages for the population served have been created by the grantees and others (Appendix 1) (Reller et al. 2008; Mahle et al. 2009; Brown et al. 2006; Liberman et al. 2014; Peterson et al. 2014; Kemper et al. 2011; Secretary's Advisory Committee on Heritable Disorders in Newborns and Children 2015).

Data Collection/Surveillance/Quality Improvement

During the project period, grantees were required to document and track the numbers of infants (1) screened, (2) with abnormal screens and referred for additional evaluations, (3) diagnosed with CCHD before discharge, and (4) diagnosed with CCHD by one year of age, but not detected via screening (i.e., false negative). Select performance characteristics of pulse oximetry screening for CCHD are summarized in Table 1. While some programs have some data on the false negatives, none of the programs has complete ascertainment of these cases. Grantees were also required to document and track the costs associated with abnormal screens and the outcomes of those detected infants (alive or deceased, number of hospitalizations, etc.). These data are not reported. Many grantees collected additional data specific to the goals and objectives of their individual projects that will be used to evaluate their screening projects (Table 2). Table 2 also describes various data collection mechanisms; some grantees used more than one mechanism for each data element, and the mechanisms varied among grantees (Table 3). The diversity of data collection and reporting methods presents a challenge to comparing data across programs, yet highlights the potential for future collaborative efforts. Table 2 also describes the many lessons learned and numerous ongoing challenges with data collection and reporting. The most common and important challenge is the lack of sustained funding for data collection activities. Some grantees have either ceased data collection activities or significantly reduced the data elements collected when grant funding ended. Further, birth certificates and/or dried blood spot cards often have limited space, therefore states using this method of collection may be restricted to collecting a limited number of fields. Some projects extract data directly from electronic medical records using tools that include labor- and time-intensive spreadsheets and automated (electronic) messaging, i.e., data file transfer. Automated systems that can extract pulse oximeter data from the testing device and transmit it to the medical record are available but not yet in wide use. Building upon existing birth defects registries by adding

reporting requirements for all failed screens in addition to CCHD diagnoses, as done in NJ, can help facilitate detailed data collection (Garg et al. 2013].

All-payer-claims databases present additional opportunities to locate data on surgical and other interventions for CCHD, including types of surgery, paid amounts and lengths of hospitalizations. If individual level data collection is not feasible, aggregate data can be collected to assess screening coverage and the number of failed screens. Data reporting and tracking may be hampered due to patient mobility, at times across state borders, for surgical and long-term follow-up care. To achieve adequate tracking, stakeholders recommended a memorandum of understanding between border state programs.

In all newborn screening programs, the identification of false negatives is a challenge. However CCHD diagnosis is a reportable condition in many states. Thus, data from birth defects registries and CCHD screening can be linked to help identify false negative screens. Programs can seek further ascertainment of false negatives through collaboration with tertiary care centers. Data collection can be instrumental for quality improvement through state level assessment and feedback of hospital performance and internal hospital audits to monitor missed screens and misinterpretation of the screening algorithm.

Telemedicine and Rural Hospitals

Newborns with a positive pulse oximetry screen and no other apparent causes of low blood oxygen saturation should have a CCHD diagnosis excluded or confirmed with clinical evaluation and echocardiography. However, echocardiography is not immediately available in all birthing locations, and many states have large geographic areas with limited access to cardiologists. Options for this scenario include echocardiography by telemedicine, mobile echocardiography units, where qualified professionals and equipment travel to the patient, or transferring to another center for evaluation, which adds significantly to the cost of screening and parental anxiety. While the infant may be initially transferred to a hospital with echocardiography capabilities, transfer to a larger hospital with a pediatric cardiac surgeon may be advised to avoid costly delays before appropriate intervention, if needed. The Wisconsin Screening Hearts In NEwborns (SHINE) Project has developed an online course for adult cardiac sonographers who might be called upon to perform neonatal echocardiography (Appendix 1) (Secretary's Advisory Committee on Heritable Disorders in Newborns and Children 2015). This allows providers who complete this training to perform this study on-site, with images read via telemedicine by a pediatric cardiologist. The decision to transport a newborn for further evaluation should be informed by a pediatric cardiology consultation.

Home Births

In 2012, over 35,000 births occurred at home in the U.S, representing almost 1% of the U.S. births (Martin et al. 2012). This population has been shown to have more than ten times the incidence of missed CCHD diagnoses than the general population (Ng and Hokansan 2010). This is likely the result of lower use of prenatal ultrasound, shorter postdelivery observation, higher incidences of certain CCHD conditions in populations that are more likely to choose home births (e.g., Amish), and a general reluctance to seek medical care. A recent

publication from one of the grantees has shown that CCHD screening can be successfully implemented in out-of-hospital births (Lhost et al. 2014). Educating midwives and lay birth attendants and providing them with pulse oximeters may improve the utilization of CCHD screening in home births. An additional challenge raised was that of outreach to unregulated/unlicensed midwives. Potential solutions are to identify a champion for midwives, provide training on CCHD screening, offer small community service-oriented grants for equipment, and make pulse oximeters available to them.

Neonatal Intensive Care Unit (NICU)

The Secretary's recommendation to add CCHD to the RUSP is targeted toward newborns in the well-baby nursery and many states are required to screen all infants, including those in the NICU. Currently, there are no recommended algorithms for NICU populations, though one has been examined (Lakshminrusimha et al. 2012). Issues complicating CCHD screening in the NICU include: receipt of oxygen; ranges of gestational age; routine use of pulse oximetry, and type and severity of disease processes, all of which impact the validity of screening. Multiple grantees report differences in NICU screening practices and currently, there are no population-based data to support CCHD screening guidelines for NICU populations. Preliminary data on the NICU population from Wisconsin and New Jersey suggest that the number of failed screens may be minimal. A collaborative effort among five states, led by NJ, is underway to evaluate and inform best screening practices in the NICU by examining timing of screening, screening values, echocardiograms conducted in response to screening results, along with clinical characteristics, such as presence of a prenatal diagnosis and/or echocardiogram prior to the screen.

Discussion

The six HRSA grantees have identified barriers and solutions to CCHD screening implementation and have provided leadership for the broader CCHD community on how to develop networks among public and private health professionals, professional organizations, family advocates, and other stakeholders to implement and improve CCHD screening programs. As the number of states engaging in CCHD screening has increased dramatically since 2011 (Glidewell et al. 2015), potential solutions to the challenges expressed by the grantees and other state representatives may aid programs to fully implement CCHD screening and to engage in quality improvement activities. The insights provided in this report reflect the experiences of six grantees and complement the significant contributions to our current understanding of CCHD screening implementation made thus far by other organizations and programs (Appendix 1) (Reller et al. 2008; Mahle et al. 2009; Brown et al. 2006; Liberman et al. 2014; Peterson et al. 2014; Kemper et al. 2011; Secretary's Advisory Committee on Heritable Disorders in Newborns and Children 2015).

Implementation and oversight for state-wide CCHD screening resulting from legislation often resides within a state public health agency. Successful implementation of legislatively mandated CCHD screening requires establishment of strong collaboration and ongoing and frequent communication between departments of health, birthing facilities, providers and

advocacy groups. The greatest benefit has been realized when engagement begins early in the implementation process such that feedback is effectively integrated.

State newborn screening (NBS) and birth defects surveillance programs are uniquely positioned to play key roles in the implementation, maintenance, and on-going assessment of CCHD screening. These two long-standing programs provide potential infrastructure on which to build CCHD screening activities. Nevertheless, birth defects surveillance programs do not exist in all states and funding to support additional activities does not often accompany legislation. One of the challenges in implementing CCHD screening and evaluation of the screening is that in many states the NBS program and birth defect surveillance programs are separate, and may have variable integration. Fostering communication and collaboration between these two stakeholders is an important challenge to consider. Further challenges to implementing CCHD screening include: obtaining resources to provide education and training, as well as to collect, store, and analyze screening data, evaluate missed cases and/or screens (false negative screens or reasons not screened), assess burden and accuracy for failed screens and resulting final diagnoses (CCHD, other cardiac, and non-cardiac), provide long-term follow-up for comprehensive outcomes, and provide data reporting/feed- back to the clinical sites (Olney et al. 2012). Implementation efforts may be limited and sustainability may be a challenge if screening mandates are unfunded. Coordinated sustainability planning can occur with collaboration among third party insurers, local and national funding sources, advocacy groups and state/ local health programs, as well as exploration of unconventional funding mechanisms (e.g. tobacco tax).

Given varied resources for data collection, the collective grantee experience provides guidance on the public health utility of various data elements using a number of collection mechanisms. These mechanisms, the majority of which were supported solely through grant funding, range from aggregate to individual level data collection with elements spanning from, for example, dichotomous pass/fail results to all potential pre- and post-ductal saturations. The benefits and limitations related to the mechanisms and questions that can be answered vary and are heavily dependent on existing infrastructure and additional funding. Accurate and complete data collection was noted by grantees to be enhanced by manual or automated quality control activities at the hospital level.

The grantees' experiences highlighted topics in CCHD screening programs that require further evaluation. Interpretation and potential modification of the screening algorithm needs further study to determine whether both the differential in saturations, as well as the less than 95% thresholds, are necessary. Data from Minnesota suggest that even minor modifications in the algorithm can have significant impact on re-screening rates (Kochilas et al. 2015). Assessment of the efficacy of CCHD screening including the magnitude of false negatives is needed along with methods for ascertaining these cases. Given the challenges in screening special populations, the most appropriate practices and algorithms for screening home births, NICU, and rural births still need to be thoroughly examined. Finally, additional provider training is needed to ensure appropriate interpretation and implementation of the screening algorithm. Ongoing education will be needed to disseminate new knowledge and evidence as they are discovered.

Early identification of CCHD is an important clinical and public health issue. CCHD detection utilizing pulse oximeter screening may lead to decreased hospital costs and infant morbidity and mortality. While costs and cost- effectiveness were not addressed as a collaborative effort, there is evidence that screening is cost-effective under a range of circumstances [Peterson et al. 2013a, 2014, 2013b]. Continued and expanded surveillance, research, prevention and communication efforts are needed to inform screening programs, with an aim to reduce morbidity, mortality and other adverse consequences for individuals and families affected by CCHD.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

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Significance

This article provides points to consider when implementing critical congenital heart disease newborn screening programs. It also provides resources and potential solutions to challenges experienced by other programs.

Critical Congenital Heart Disease Screening Table

Foot Application Sites RH Application Sites
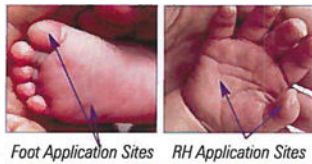
Right Hand	Either Foot											<90
100	100	99	98	97	96	95	94	93	92	91	90	<90
99	100	99	98	97	96	95	94	93	92	91	90	<90
98	100	99	98	97	96	95	94	93	92	91	90	<90
97	100	99	98	97	96	95	94	93	92	91	90	<90
96	100	99	98	97	96	95	94	93	92	91	90	<90
95	100	99	98	97	96	95	94	93	92	91	90	<90
94	100	99	98	97	96	95	94	93	92	91	90	<90
93	100	99	98	97	96	95	94	93	92	91	90	<90
92	100	99	98	97	96	95	94	93	92	91	90	<90
91	100	99	98	97	96	95	94	93	92	91	90	<90
90	100	99	98	97	96	95	94	93	92	91	90	<90
<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90	<90

PASS – Pulse ox of 95% or more in RH or foot AND difference of 3% or less between the two.
Action: Do not repeat for screening. Provide normal newborn care.

RETEST – Pulse ox of 90-94% in BOTH the RH and foot OR a difference of 4% or more between the RH and foot.
Action: Repeat pulse ox measurements in one hour.
If the second test remains yellow, repeat again in one hour. If the third test is still yellow, it is a FAIL and should be reported to the medical provider

FAIL – Pulse ox of 89% or less in either the RH or foot.
Action: Do not repeat, report to medical provider.

Adapted from the Mueller CCHD Screening Chart created by Cynthia Mueller RN, BSN (2013)
Adapted from *Strategies for Implementing Screening for Critical Congenital Heart Disease*
Kemper A., et al., Pediatrics 2011(128)5: e1259 - 1267.Kemper A., et al., Pediatrics 2011(128)5: e1259 - 1267.

VDH VIRGINIA DEPARTMENT OF HEALTH NEWBORN SCREENING UNIVERSITY of VIRGINIA Children's Hospital

Fig. 1.
A visual tool developed by the state of Virginia to enhance CCHD screening algorithm interpretation

Table 1

Critical congenital heart disease newborn screening data collection by six grantees

State/program	Dates of data collection	# of Babies screened	# Screen positive	Positive rate (per 10,000)	# of CCHD Diagnoses	Rate of diagnosis (per 10,000)
Michigan	April 2014–May 2015	98,158	35	3.6	4[a]	0.4
New England[b]	Varies	48,294	27	5.6	3[a]	0.6
New Jersey[c]	August 2011–December 2015	427,216	266	6.2	20[a]	0.5
Utah[d]	May 2013–September 2014	10,662	54	50.6	1[a]	0.9
Virginia[e]	January 2014–June 2015	68,479	200	29.2	23[f]	3.4
Wisconsin	January 2013–December 2014	55,329	63	11.4	7[a]	1.3
Total		708,318	645	11.4 (95% CI 5.1–25.2)	58	0.9 (95% CI 0.4–2.3)

[a] CCHD common truncus, transposition of great vessels, tetralogy of Fallot, pulmonary atresia, tricuspid atresia, hypoplastic left heart syndrome, total anomalous pulmonary venous connection, coartation of aorta, double outlet right ventricle, Ebstein anomaly, interrupted aortic arch, single (common) ventricle

[b] Maine, New Hampshire, Vermont, Rhode Island and Connecticut; Data collected from eight institutions over varying time periods from June 2012–May 2015

[c] NJ legislation mandates screening of all infants including those with prenatal diagnosis of CHD and/or echocardiogram prior to screening. This number includes all fails reported to the NJ Birth Defects Registry during the time period specified

[d] Data collected from two institutions

[e] The screen positive number includes misinterpretations of the screening algorithm. The number that actually failed all three screens is not known. The number of CCHD diagnoses includes all diagnoses, not only those detected via screening

[f] CCHD = common truncus, transposition of great vessels, tetralogy of Fallot, pulmonary atresia, tricuspid atresia, hypoplastic left heart syndrome, total anomalous pulmonary venous connection

Table 2

Data and collection mechanism useful in evaluation of CCHD screening by six demonstration project grantees

Data collected by grantees	Collection mechanisms	Area of evaluation
Population characteristics		
Number live-births	EBC, AG	Screening coverage
Number screened	AG, EMR, HL7, SEM, EBC, BDR, NBC	
Reason not screened	AG, EMR, HL7, SEM, EBC, BDR, NBC	
Screening results		
Pulse oximetry screen date/time	EMR, HL7, SEM, EBC, BDR, NBC	Screening algorithm performance and interpretation
All saturation results	EMR, HL7, SEM, EBC, BDR	
Pass/Fail	AG, EMR, HL7, SEM, EBC, BDR, NBC	
Clinical characteristics		
Antenatal ultrasound (yes/no)	EMR, HL7, SEM, EBC, BDR	Screening algorithm performance
Prenatal CCHD diagnosis	EMR, HL7, SEM, EBC, BDR	Contribution of pulse oximetry screening to identification of CCHD
Clinical exam date/time	EMR, HL7, SEM, EBC, BDR	
Clinical exam findings	EMR, HL7, SEM, EBC, BDR	
Echocardiogram (yes/no)	EMR, HL7, SEM, EBC, BDR	
Echocardiogram date/location or prior to screen	EMR, SEM, EBC, BDR	
Echocardiogram findings	EMR, SEM, EBC, BDR	
Signs or symptoms at the time of screen	EMR, SEM, EBC, BDR	
Final diagnosis	EMR, SEM, EBC, BDR	
Age at diagnosis	EMR, EBC, BDR	
Follow-up information		
Surgery date/hospital	EMR, EBC, BDR	Outcomes
Date of death (if applicable)	BDR, Vital Records	

NBC newborn bloodspot card, *SPF* state-specific CCHD paper forms, *SEM* state-specific CCHD electronic module, *HL7* Health level-7 messaging; automatic file transfer, *AG* aggregate data collection, *EBC* electronic birth certificate, *BDR* birth defects registry, *EMR* hospital electronic medical record

Table 3

Data collection challenges and lessons learned, by six CCHD demonstration project grantees

Description of grantees' data collection activities with demonstration grant funding	Lessons learned	Continuing challenges

Michigan
    Funded mandate for screening
    Data incorporated into newborn screening Laboratory Information Management System
    Data collection mechanism: EBC, EMR, HL7, SEM, BDR
    Individual level data collected
	Support needed from birthing facilities, IT departments
	Internal quality control (QC) audits needed at hospital Communication and support from birthing facilities are crucial to identify and follow-up on failed, missed and transferred cases level
		Hospital readiness for HL7 submission
		Data quality: file transfer, completeness, and follow-up on missed or failed screens
		Obtaining internal and external data use agreements to evaluate false negatives

New England (ME, NH, VT, RI, CT)
    NH and CT do not have public health authority for data collection
    Project collected de-identified data from eight birthing facilities across five states (two aggregate and six individual-level)
    Data collection mechanism: AG, SPF, SEM, BDR
	Screening at tertiary care centers has a very low detection rate, likely due to prenatal diagnosis and clinical diagnosis post-natally prior to screening
	False positive rate is acceptably low Public health programs and birthing facilities worked cooperatively across states and shared resources, expertise and experiences
		Data are not collected consistently across states and are reported in different formats
		Algorithm interpretation errors
		Home birth and NICU screening data not collected

New Jersey
    Unfunded mandate for screening
    As of January 2015, individual level data incorporated into electronic birth record system
    Failed screens reported to NJ Birth Defects Registry (BDR)
    Data collection mechanism: AG, EBC, BDR
	Internal quality control (QC) audits needed at hospital level
	Strong communication and support from birthing facilities required for implementation
	Lack of real time reporting and inability to track infants across reporting periods with aggregate data collection
	Individual level reporting improves ability to conduct QC and evaluate false negatives
		Identification of false negatives due to tertiary care provided in bordering states
		No direct data input from EMR into electronic birth certificate and manual data entry required for individual level reporting
		No real-time QC of algorithm interpretation

Utah
    Unfunded mandate for screening
    BDR reporting rule
    One year of pilot data with individual saturation levels from two large hospital systems
    As of October 2014, using EBC to monitor pass/fail
    Link EBC to BDR to confirm true fails and identify false fails
    Data collection mechanism: AG, EBC, EMR, BDR
	Cumbersome for birthing facilities to incorporate screening results into their current EMR
	Difficulty in data cleaning
	EBC reliable for Pass/Fail results and not onerous for facilities
	Need to work with legislature to include mandated reporting and funding
		Reporting errors by EBC clerks
		Timing of EBC reporting does not include NICU results on most babies
		Working with home birth providers to screen/report
		Algorithm interpretation errors

Virginia
    January 2014 individual level data incorporated into EBC
    CCHD diagnosis reported into BDR
    Legislation mandates screening and reporting for birth hospitals
    Data collection mechanism: AG, EBC, EMR, BDR
	Failure mode and effects analysis (FMEA) study testing AAP algorithm improved screening processes at each hospital
	Majority of reported failed screens were documentation errors.
	QC activities include clarifying language in EBC for more accurate reporting
	Cross referencing EBC and BDR is essential to identify children with CCHD. Most children with CCHD reported to BDR were missing screening results in EBC. Incomplete screening results noted for prenatally diagnosed infants
	Communication and education need for birth registrars as well as providers in birth hospitals
	Hospital site visits beneficial for QC and education
		Staff turnover in birthing hospitals specifically EBC registrars
		Direct report of results into EBC, but no automatic algorithm interpretation Registrars required to interpret and document actual result
		Due to lack of funding, limited DOH staff to conduct follow-up and referral
		Reporting compliance
		Missed screens for specific populations
		Legislation does not mandate screening in NICUs or for midwives
		Continued funding needed to support educational website

Wisconsin
    CCHD screening and clinical data linked to newborn hearing screening surveillance
    Newborn blood spot card used as the data collection tool for CCHD Minimum Data Set for all infants
	CCHD screening in the home birth community was well received coincident with an increased newborn blood screening rate
	Mechanism to document reason for not screening was needed to allow hospitals to document compliance
	Misinterpretation of screening algorithm educational materials were beneficial
		Substantial manual data entry is required to enter the Minimum
		Data Set and to collect and enter the Expanded Data Set
		Reporting rates continue to vary significantly between hospitals; ongoing education and QC are required

Description of grantees' data collection activities with demonstration grant funding	Lessons learned	Continuing challenges
Expanded Data Set collected on infants who fail CCHD screening or are found to have CCHD Data collection mechanism: EBC, NBC, SPF, SEM	Coordination of newborn blood, hearing, and CCHD screening education, data collection, and surveillance important Hospital specific reporting rates required to evaluate implementation and effectiveness of screening	Screening evaluation ends at the time of diagnosis and no statewide evaluation of CCHD treatment is available

AG aggregate data collection, *EBC* electronic birth certificate, *BDR* birth defects registry, *EMR* hospital electronic medical record, *NBC* newborn bloodspot card, *SPF* State-specific CCHD paper forms, *SEM* State-specific CCHD electronic module, HL7 Health level-7 messaging; automatic file transfer