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Strategies for Implementing Screening for Critical Congenital Heart Disease

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KEY WORDS

congenital heart defects, neonatal screening, oximetry

ABBREVIATIONS

HHS—US Department of Health and Human Services
SAGHDNC—Secretary's Advisory Committee on Heritable Disorders in Newborns and Children
AAP—American Academy of Pediatrics
AHA—American Heart Association
HRSA—Health Resources and Services Administration
CCHD—critical congenital heart disease
ACCF—American College of Cardiology Foundation
FDA—US Food and Drug Administration

The findings and conclusions in this report are those of the authors and do not necessarily represent the views of the Centers for Disease Control and Prevention, the Health Resources and Services Administration, the National Institutes of Health, the Food and Drug Administration, or the US Department of Health and Human Services.

This report has been endorsed by the American Academy of Pediatrics, the American College of Cardiology Foundation, and the American Heart Association Council on Cardiovascular Disease in the Young.

(Continued on last page)

abstract

FREE

BACKGROUND: Although newborn screening for critical congenital heart disease (CCHD) was recommended by the US Health and Human Services Secretary's Advisory Committee on Heritable Disorders in Newborns and Children to promote early detection, it was deemed by the Secretary of the HHS as not ready for adoption pending an implementation plan from HHS agencies.

OBJECTIVE: To develop strategies for the implementation of safe, effective, and efficient screening.

METHODS: A work group was convened with members selected by the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children, the American Academy of Pediatrics, the American College of Cardiology Foundation, and the American Heart Association.

RESULTS: On the basis of published and unpublished data, the work group made recommendations for a standardized approach to screening and diagnostic follow-up. Key issues for future research and evaluation were identified.

CONCLUSIONS: The work-group members found sufficient evidence to begin screening for low blood oxygen saturation through the use of pulse-oximetry monitoring to detect CCHD in well-infant and intermediate care nurseries. Research is needed regarding screening in special populations (eg, at high altitude) and to evaluate service infrastructure and delivery strategies (eg, telemedicine) for nurseries without on-site echocardiography. Public health agencies will have an important role in quality assurance and surveillance. Central to the effectiveness of screening will be the development of a national technical assistance center to coordinate implementation and evaluation of newborn screening for CCHD. *Pediatrics* 2011;128:e1259–e1267

Newborn screening has led to dramatic improvements in morbidity and mortality rates for a variety of conditions.¹ Historically, newborn screening has been based on analysis of dried blood spots and has operated as a partnership between health care providers, who obtain the samples and oversee medical follow-up, and state-based public health systems, which analyze the dried blood spots, assist health care providers and families in follow-up, and monitor the effectiveness of the screening process through surveillance activities. The US Health and Human Services (HHS) Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) was authorized by the US Congress to provide guidance to the Secretary of the HHS about which conditions should be included in newborn screening and how systems should be developed to ensure appropriate screening and follow-up care.^{2,3}

Before 2010, the only condition recommended for newborn screening that did not follow the dried-blood-spot paradigm was newborn hearing screening. Newborn hearing screening relies on in-hospital testing before discharge and subsequent outpatient audiology testing for those with abnormal results.⁴ Unlike dried-blood-spot testing, individual hospitals and birthing centers had to invest in screening devices, maintain sufficient numbers of skilled staff to conduct the screening and interpret the results, and develop systems to track and communicate results of testing with public health departments, health care providers, and families. Because results of hearing screening originate in the hospitals and birthing centers, public health programs face significant challenges to ensuring follow-up to ensure the success of newborn hearing screening.^{5,6}

In September 2010, the SACHDNC recommended that critical congenital cy-

anotic heart disease be added to the recommended uniform screening panel on the basis of findings from a comprehensive evidence review. The goal of this recommendation was to identify those newborns with structural heart defects usually associated with hypoxia in the newborn period that could have significant morbidity or mortality early in life with closing of the ductus arteriosus or other physiologic changes early in life. The SACHDNC considered 7 specific lesions as primary targets for screening on the basis of advice from a technical expert panel: hypoplastic left heart syndrome; pulmonary atresia; tetralogy of Fallot; total anomalous pulmonary venous return; transposition of the great arteries; tricuspid atresia; and truncus arteriosus. This subset of lesions excludes those not usually associated with hypoxia (eg, aortic valve stenosis).⁷

This recommendation built on a 2009 statement from the American Academy of Pediatrics (AAP) and the American Heart Association (AHA), which found compelling reasons for newborn screening but called for "studies in larger populations and across a broad range of newborn delivery systems" before pulse-oximetry screening should be recommended.⁷ The SACHDNC was especially persuaded by a prospective screening study of nearly 40 000 newborns in Sweden⁸ and a separate study of nearly 40 000 newborns in Germany.⁹ Comparing the accuracy of pulse-oximetry monitoring for the 7 defects specified by the SACHDNC to that of these other studies was somewhat challenging because of differences in the lesions that were targeted for detection by the screening. For example, the study in Sweden considered all ductal-dependent lesions. The researchers' approach, for example, was to add critical aortic stenosis and coarctation of the aorta but

exclude tetralogy of Fallot. With this case definition, the study from Sweden found the sensitivity of pulse-oximetry monitoring to be 62.1% and the specificity to be 99.8%; the false-positive rate was 0.17%. In contrast, the AAP/AHA statement used a broader definition, which included all lesions that would require surgery or catheter intervention in the first year of life.

The SACHDNC made the recommendation for screening with the understanding that specific activities would be undertaken, including having the Health Resources and Services Administration (HRSA) guide the development of screening standards and the infrastructure needed for implementation of a public health approach to point-of-service screening and developing education materials; having research conducted by the National Institutes of Health; and surveillance and tracking by the Centers for Disease Control and Prevention. However, the Secretary of the HHS did not endorse the recommendation from the SACHDNC to begin screening, in part because of questions about how to implement that screening. Some states (eg, Maryland, New Jersey) have legislation that promotes newborn screening for critical congenital heart disease (CCHD), which increases the urgency for a draft implementation plan.

The SACHDNC, in collaboration with the AAP, the American College of Cardiology Foundation (ACCF), and the AHA, convened a work group to outline implementation strategies for the SACHDNC, which are summarized here. It is important to recognize that many newborns with the targeted congenital heart defects do not develop clinically appreciable cyanosis until after nursery discharge, and some lesions (eg, hypoplastic left heart syndrome) may present with significant cardiovascular compromise without apparent cya-

nosis. Therefore, the work group recommended renaming the target conditions "critical congenital heart disease" (CCHD) (omitting the word "cyanotic").

METHODS

A work group was convened for a 2-day meeting in January 2011. Work-group members (see Appendix) included primary care providers; specialists, including pediatric cardiologists and neonatologists; nurses; representatives from the AAP, the ACCF, the AHA, the American College of Medical Genetics, the March of Dimes, the Association of Maternal and Child Health Programs, the Association of Public Health Laboratories, and the SACHDNC; parent screening advocates; state public health officials; and representatives from the Centers for Disease Control and Prevention, the US Food and Drug Administration (FDA), the HRSA, and the National Institutes of Health. Included were people who have implemented pulse-oximetry monitoring for CCHD in newborn nurseries in Arkansas, California, Minnesota, New York, Washington, and Washington, DC. The work group was moderated by William T. Mahle, MD, a pediatric cardiologist who led the development of the 2009 AAP/AHA statement,⁷ and R. Rodney Howell, MD, chair of the SACHDNC. The work group was supported by other invited experts, including those from the Centers for Disease Control and Prevention and the FDA, and 2 who had conducted large-scale studies of screening in Europe. The work-group meeting was open to the public.

The meeting focused on recommendations for pulse-oximetry monitoring for CCHD, including recommendations for the service infrastructure needs for follow-up, and strategies for filling in important knowledge gaps. A smaller writing group prepared a summary report of the meeting, which

was then iteratively revised with the work group until agreement was obtained. The report was subsequently reviewed by the AAP, the ACCF, and the AHA, each of which endorsed this report.

RESULTS

Screening Population and Targets

The work group chose to focus initially on screening in the well-infant nursery because of the risk of missed cases of CCHD among healthy-appearing newborns. The work group recognized the importance of also considering screening within NICUs. However, developing a simple algorithm for the NICU setting is challenging because of the heterogeneity of underlying conditions (eg, prematurity, meconium-aspiration syndrome, sepsis). Unlike the well-infant nursery, many infants in the NICU undergo repeated medical evaluations, are monitored by pulse oximetry, and have longer lengths of stay. However, there was concern that screening only in well-infant nurseries would miss newborns with short stays in intermediate care nurseries. The work group endorsed screening infants in intermediate care nurseries or other units in which discharge is common in the first week by using the work-group protocol for screening in the well-infant nursery. The work group chose not to focus on out-of-hospital births, which raise challenging coordination-of-care issues, which will be addressed in the future.

One of the advantages of pulse-oximetry monitoring is the ability to detect other hypoxic cardiac- or non-cardiac-associated conditions (eg, persistent pulmonary hypertension), characterized by the SACHDNC as targets secondarily detected by the screening technology ("secondary targets"). Secondary targets are common to other newborn screening tests (eg,

identification of hemoglobin H disease when screening for sickle cell anemia¹⁰). Although the primary goal of screening on the basis of the SACHDNC recommendation is identification of the 7 specific lesions associated with CCHD, tracking rates of identification of important secondary targets could lead to modifications of the screening protocol.

Screening Technology

The work group recommended that screening be performed with motion-tolerant pulse oximeters¹¹ that report functional oxygen saturation, have been validated in low-perfusion conditions, have been cleared by the FDA for use in newborns, and have a 2% root-mean-square accuracy. Commercially available pulse oximeters often are labeled by manufacturers according to generation of technology (eg, "next generation"). However, generation designation is not standardized and may not be related to validity or reliability. Furthermore, no standards have been developed regarding motion tolerance. A new guidance document on the safety and effectiveness of pulse oximeters is being developed by the FDA.¹² When the guidance document is finalized, any pulse oximeter used for screening should meet FDA recommendations. Having specific FDA-cleared labeling and conformance to the relevant standard¹³ will be an important strategy for ensuring that appropriate devices are used for screening.

Pulse oximeters can be used with either disposable or reusable probes. Reusable probes can reduce the cost of screening, but they must be appropriately cleaned between uses to minimize the risk of infection. Some probes have been developed to be partially reusable, which reduces the need to clean between uses and are less expensive than fully disposable

probes. Probes with close coupling to skin (ie, taped rather than clamped) provide better performance for oximetry monitoring in newborns. Pulse oximeters are validated only with the specific probes recommended by the manufacturer; therefore, to optimize valid screening, manufacturer-recommended pulse-oximeter-probe combinations should be used.

Screening Criteria

The work group recommended that screening not begin until 24 hours of life, or as late as possible if earlier discharge is planned, and be completed on the second day of life. Earlier screening can lead to false-positive results because of the transition from fetal to neonatal circulation and stabilization of systemic oxygen saturation levels, and later screening can miss an opportunity for intervention before closing of the ductus arteriosus. Screening was recommended in the right hand and 1 foot either in parallel or in direct sequence. The pulse-oximetry measure is complete once the waveform on the oximeter's plethysmograph is stable or there is another indication that the device is appropriately tracking the infant's pulse rate.

Selecting the threshold for a positive pulse-oximetry monitoring result is challenging, because it must trade-off the harm of missing CCHD against the harm of false-positive screen results. None of the studies reviewed by the SACHDNC included receiver operator characteristic curves developed from primary data, which would allow a direct evaluation of this trade-off. However, on the basis of new data from the large population-based screening activities in Sweden⁹ and England,¹⁴ the work group developed a recommendation for screening that was based on what was shown to be effective in those studies.

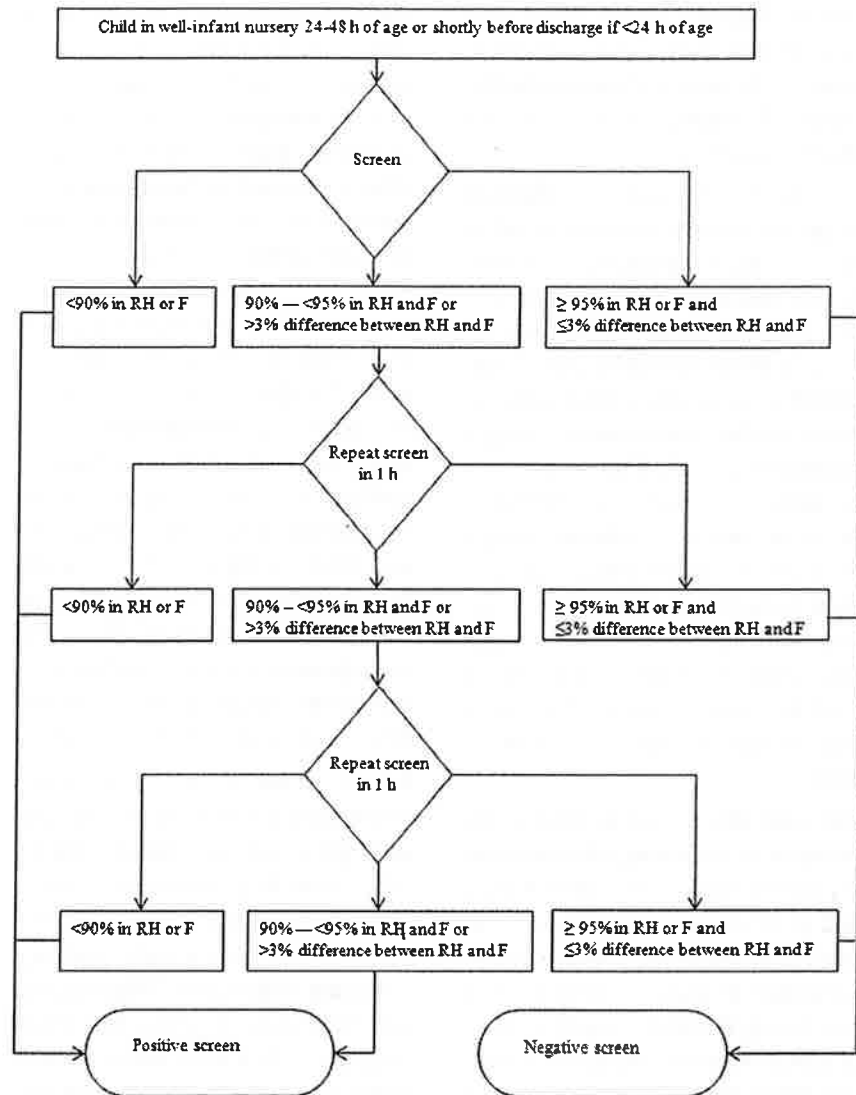


FIGURE 1

The proposed pulse-oximetry monitoring protocol based on results from the right hand (RH) and either foot (F).

The screening protocol is listed in Fig 1. A screen result would be considered positive if (1) any oxygen saturation measure is $<90\%$, (2) oxygen saturation is $<95\%$ in both extremities on 3 measures, each separated by 1 hour, or (3) there is a $>3\%$ absolute difference in oxygen saturation between the right hand and foot on 3 measures, each separated by 1 hour. Any screening that is $\geq 95\%$ in either extremity with $\leq 3\%$ absolute difference in oxygen saturation between the upper and lower extremity would be considered a

“pass” result, and screening would end.

Anecdotal reports have suggested that false-positive results are decreased if the infant is alert, possibly by reducing the likelihood of low oxygen saturations caused by hypoventilation in deep sleep. In addition, timing pulse-oximetry monitoring around the time of the newborn hearing screening improves efficiency, assuming that the hearing screening is conducted after 24 hours or immediately before dis-

charge. The particular screening strategy should reflect the conditions within each particular nursery and the needs of infants, families, and the health care providers.

The work group noted that performing a typical physical examination alone for CCHD led to almost 10 times more false-positive results compared with using similar screening protocols in Sweden and the United Kingdom.^{8,14} Repeated pulse-oximetry testing after an initial positive screen result if oxygen saturation is <95% in both extremities or there is a >3% absolute difference in oxygen saturation between the right hand and foot, as illustrated in the protocol, lowers the likelihood of a false-positive result compared with a single measurement. However, there is no need to repeat pulse-oximetry testing if the oxygen saturation is <90% in any screen.

The work group emphasized the importance of not having pulse-oximetry monitoring replace a complete history and physical examination, which can sometimes detect CCHD before the development of hypoxia. Pulse-oximetry monitoring, therefore, should be used to complement the physical examination. Although agreement was reached on the screening protocol, the work group was concerned that this screening protocol might lead to high rates of false-positive results in high-elevation communities, such as those in Denver, Colorado.^{15–17} The criteria for a positive screen result may need to be modified for these areas. Regardless of the specific screening thresholds, comprehensive training will be central to implementing safe and effective screening.

Diagnostic Strategies

Any newborn with a positive screen result first requires a comprehensive evaluation for causes of hypoxemia. In the absence of other findings to ex-

plain hypoxemia, CCHD needs to be excluded on the basis of a diagnostic echocardiogram (which would involve an echocardiogram within the hospital or birthing center or transport to another institution) or through the use of telemedicine for remote evaluation. The work group also emphasized the need for high-quality echocardiograms with interpretation by a pediatric cardiologist because of the challenge of diagnosis in some cases (eg, total anomalous pulmonary venous return). The work group recommended against replacing a diagnostic echocardiogram with other evaluations (eg, chest radiograph, electrocardiogram, hyperoxia test), which can be inaccurate for diagnosing CCHD. The work group endorsed consulting a pediatric cardiologist, when feasible, before obtaining the echocardiogram.

Because of the importance of quickly establishing the diagnosis of CCHD, the work group recommended that hospitals and birthing centers establish a protocol to ensure timely evaluation, including echocardiograms and any necessary subsequent follow-up, before instituting a CCHD screening program. Future work will be needed to ensure the quality of in-center and telemedicine approaches to echocardiography. The work group also recognized the importance of training an adequate number of pediatric cardiologists to ensure that diagnostic services are available on-site, with short-distance transport, or through telemedicine. Similarly, pediatric cardiac surgery centers will have to be prepared to accept newborns with CCHD identified by pulse oximetry.

Connection to the Medical Home

The results of newborn CCHD screening should be communicated to newborns' primary care providers. During the first outpatient visit, primary care

providers should ensure that all newborns were appropriately screened and received any necessary follow-up. The work group recognized the importance of developing health information exchange systems to allow primary care providers, in addition to cardiology subspecialists, to easily track this information. To facilitate this tracking, standards for electronic reporting of pulse-oximetry measurements will need to be developed. Standards for electronic reporting would also help facilitate the development of quality measures.

Primary care providers will also need to develop strategies for screening those newborns who missed screening. As with other newborn screening tests, primary care providers play a central role in ensuring long-term follow-up for those infants diagnosed with CCHD through newborn screening and coordinating their care with a pediatric cardiologist.²

Public Health, Quality Assurance, and Surveillance

Follow-up for a positive screen result should be managed by the hospital or birth center before discharge; therefore, the role of public health agencies in CCHD screening is different from that in the case of newborn dried-blood-spot screening or newborn hearing screening. However, public health agencies can play a central role in quality assurance and surveillance. There are several challenges to public health agencies' involvement with CCHD screening, including the inability to collect real-time screening data through health information exchange systems, absence of the direct presence of public health personnel in hospitals and birthing centers, and the financial and staffing pressures within public health departments.

State-level Title V Maternal and Child Health programs and birth-defect sur-

veillance and prevention programs should play a role in surveillance and evaluation of CCHD screening. These programs already conduct public education and outreach; train providers; and support genetic services, newborn screening programs, and services for children with special health care needs. Although state birth-defect programs could assist with CCHD surveillance, there are differences across states in resources for such activities and the approaches to case ascertainment. As of February 2011, there were 40 birth-defect surveillance programs in the United States and 6 more in development. With adequate resources, some of these programs could potentially collect and track data on populations screened or not screened or those with false-negative screening results. Data could also be collected on whether a diagnosed CCHD was detected through prenatal ultrasound or newborn pulse-oximetry monitoring. Collecting data to understand the factors associated with false-positive pulse-oximetry monitoring results could also help refine the recommended screening activities. Although there is currently no capacity in birth-defect programs to undertake real-time follow-up of CCHD-positive screen results, including short-term follow-up, the infrastructure is in place in many states for birth-defect surveillance programs to play a critical role in conducting long-term surveillance and evaluation.

Health Care Costs

The main costs of a screening program for CCHD are related to staff time for screening, tracking results, and communicating with parents, the purchase and maintenance of screening equipment, consumables associated with screening (eg, probes, adhesive wraps, cleaning supplies), the costs associated with verifying a positive screen result, and the costs associ-

ated with treatment. The cost of conducting pulse-oximetry examination and follow-up is quite low in absolute terms; published estimates are \$5 or less per infant^{7,8} up to \$10 per infant, depending on the protocol.¹⁴ Although screening can sometimes be completed in <1 minute, other studies have estimated that the process takes 5 minutes of staff time, including communication with parents.¹⁴ The cost estimate compares quite favorably with cost estimates for newborn hearing screening (\$30 or more per infant with an average reimbursement by private health plans in 2004 of \$84 if billed separately¹⁷). Moreover, the cost of pulse oximetry is significantly offset by avoided costs of care. The authors of the report from Sweden calculated that the savings in health care costs from the prevention of 1 case of complications of circulatory collapse resulting from an undiagnosed CCHD may exceed the cost of screening 2000 newborns.⁹

Another potentially important cost is related to delayed discharge because of the need to repeat screening or obtain diagnostic evaluation, which leads to extra hospital days that may not be reimbursed by insurance carriers. Echocardiography is typically reimbursed well. However, the cost of transport can be high and receive variable insurance reimbursement. Although telemedicine for remote echocardiography could be important for hospitals and birthing centers without ready access, it is unclear who would pay to develop and maintain the infrastructure.

At present, there is no clear way to bill for pulse-oximetry monitoring, because the currently available *Current Procedural Terminology* (CPT) codes for pulse oximetry are only appropriate when accompanied by a diagnostic code for a pulmonary disease associated with hypoxia.¹⁹ The AAP, AHA,

and ACCF should work with the American Medical Association, which develops CPT codes, to develop the appropriate CPT codes for pulse-oximetry monitoring and with public and private payers to ensure appropriate reimbursement. However, newborn hospital-based screening services such as hearing screening are commonly not reimbursed separately if conducted by regular hospital nursery staff, even with appropriate CPT codes available. Because the cost of conducting pulse-oximetry monitoring is quite low, the cost to hospitals and birthing centers should not be a major barrier. In Switzerland, for example, most birthing centers have adopted pulse-oximetry monitoring, and an estimated 85% of infants are screened despite no mandate for either screening or insurance reimbursement for screening.²⁰

The work group recognized the concerns about limited health care resources and emphasized the need to weigh the costs of pulse oximetry against the potential benefits of early diagnosis of CCHD, including the costs saved by decreasing the morbidity associated with later diagnosis. Cost data should be compared with the screening-outcomes data, such as those collected by public health agencies, to inform policymakers and to develop new interventions to improve the efficiency of screening.

Health Care Provider and Family Education

Both health care providers and families must understand the rationale for and limitations of pulse-oximetry monitoring to detect CCHD, including the important understanding that a negative screening result does not exclude the possibility of CCHD or other congenital heart disease. Similarly, educa-

tion is needed to minimize the harm that may be generated by false-positive screen results. Implementation of other newborn screening tests has been improved through the development of simple clinical decision-support tools for health care providers that explain the screening and what should be done in the event of a positive result (eg, the HRSA-funded ACTion sheets and simple fact sheets for families).²¹ Similar materials need to be developed for pulse-oximetry monitoring and should be available in print and through electronic media in English, Spanish, and other local languages. Implementation toolkits used to help hospitals and birthing centers assess their degree of readiness for screening, to develop algorithms for screening, and to evaluate their ongoing activities are also important.

Coordination of Implementation Activities

The work group endorsed the development of a national clearinghouse and technical assistance center similar to the National Resource Center for Newborn Hearing Screening (www.infanthearing.org), the National Newborn Screening and Genetics Resource Center (<http://genes-r-us.uthscsa.edu>), and the Emergency Medical Services for Children National Resource Center (www.childrensnational.org/EMSC). These sites provide examples of ways to coordinate service delivery between health care providers and state public health agencies. Replicating this approach through partnership with state Title V Maternal and Child Health programs would allow implementation that takes into account specific local factors such as the availability of diagnostic services.

DISCUSSION

A significant body of evidence suggests that early detection of CCHD through

pulse-oximetry monitoring is an effective strategy for reducing morbidity and mortality rates in young children. The work group identified strategies for hospitals and birthing centers to implement pulse-oximetry monitoring for CCHD and included the following specific recommendations.

- Screening should be conducted by using motion-tolerant pulse oximeters that report functional oxygen saturation and have been cleared by the FDA for use in newborns.
- Screening should be based on the recommended screening algorithm and be performed by qualified personnel (eg, nurses, allied health technicians) who have been educated in the use of the algorithm and trained in pulse-oximetry monitoring of newborns.
- The algorithm cutoffs may need to be adjusted in high-altitude nurseries.
- Any abnormal pattern of low blood oxygen saturation requires a complete clinical evaluation by a licensed, independent practitioner. In the absence of other findings to explain hypoxemia, CCHD needs to be excluded on the basis of a comprehensive echocardiogram interpreted by a pediatric cardiologist before discharge from the hospital. If an echocardiogram cannot be performed in the hospital or birthing center and diagnosis by telemedicine is not possible, strong consideration should be made for transfer to another medical center for diagnosis. Before implementing screening, protocols for arranging diagnostic follow-up should be established.
- Hospitals and birthing centers should establish partnerships with local and state public health agencies to develop strategies for quality

assurance and monitor the impact of screening.

- Primary care providers should ensure that newborns in their practice were appropriately screened and should work to facilitate long-term follow-up for those diagnosed with CCHD.
- Standards should be developed for electronic reporting of pulse-oximetry monitoring and diagnostic outcomes.

CONCLUSIONS

The work group recognized the challenges of implementing a new screening program. To ensure that screening is implemented in a safe and effective manner, the work group strongly endorsed the development and funding of a national technical assistance center to disseminate best practices; to partner with public health agencies to monitor the impact of screening; to evaluate and make recommendations regarding workforce and related infrastructure needs; and to coordinate research to help answer the important unanswered questions regarding screening thresholds and optimal strategies for diagnosis and follow-up. The Secretary of the HHS has directed an interagency work group to develop a plan to address these critical gaps before recommending that CCHD be a part of the recommended uniform screening panel.

APPENDIX: WORK-GROUP MEMBERS

The following is a list of work-group members and the agencies or organizations they represented at the meeting (being listed as a work-group member does not imply that the members or the organization that they represent endorse all aspects of this report): Mona Barmash (Congenital Heart Information Network, Margate City, NJ), Robert H. Beekman, MD (Cincinnati Children's Hospital Medical Center,

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