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## A Novel, More Efficient, Staged Approach for Critical Congenital Heart Disease Screening

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### Abstract

**Objective**—Screening for critical congenital heart disease (CCHD) using pulse oximetry has been endorsed by the AAP and the AHA. The recommended screening requires two saturation readings. We sought to determine the incidence of undetected CCHD in Tennessee for the two years following implementation of an algorithm that assigned an immediate pass to a single lower extremity saturation of 97% or higher.

**Study Design**—State Genetic Screening records and reports of missed cases from the Tennessee Initiative for Perinatal Quality Care were used to determine if CCHD cases were missed by the new screening algorithm.

**Result**—During the study, 232 infants failed the screen with 51 or 22% true positives, 13 infants had undetected CCHD (10 coarctations, 2 Anomalous veins, 1 Tetralogy of Fallot).

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**Conclusion**—This approach eliminated over 150,000 pulse oximetry determinations in Tennessee without affecting the ability of pulse oximetry to detect CCHD before discharge.

### Keywords

pulse oximetry screening; screening algorithm; critical congenital heart disease; state screening; coarctation of aorta; healthcare costs

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### Introduction

In the US, about 7,200 (or 18 per 10,000) babies born each year have one of seven critical congenital heart defects (CCHDs). Historically up to 30 % of these infants were not diagnosed until after discharge or after death. [1]

In 2011, the US Secretary of Health and Human Services recommended universal screening of newborns for critical congenital heart defects using pulse oximetry. As of 2015 at least 43 states have passed a law or regulation that requires hospitals to screen newborns for critical CHDs. Many hospitals in the remaining states choose to screen newborns, even though their state does not require it. [2] The protocol recommended by the AHA and CDC for screening [3] is based on extensive review of evidence from major European trials. [4–6]

The AHA protocol evaluates the pulse oximetry reading of the infant on a hand and foot after 24 hours of age. To avoid the problem of multiple false positives it was decided to repeat the screen twice prior to declaring the infant a failure. Seven key lesions which would be expected to have a low saturation were targeted by the AHA. These included hypoplastic left heart, pulmonary atresia, Tetralogy of Fallot, total anomalous venous return, transposition of the great arteries, tricuspid atresia and truncus arteriosus.

Screening two extremities is based on the possibility that a baby with right to left ductal shunting from hypoplastic left heart or secondary targets such as aortic coarctation or ductal dependent systemic circulation may have decreased saturations in the lower extremity compared to the upper extremity, therefore a persistent difference greater than 3 % between upper and lower extremity is an indication for further evaluation. Using this protocol it was estimated that about half the infants with CCHD would be diagnosed, who would otherwise have been missed by lack of prenatal diagnosis and absence of signs in the newborn nursery. [7]

Ailes et al. [7] estimated that approximately 875 newborns with critical CHD could be identified at birth hospitals in the U.S. each year by using pulse oximetry newborn screening. An equal number (880 babies) might still be missed, including truncus arteriosus, coarctation, and interrupted aortic arch. These lesions are less likely to be detected through pulse oximetry newborn screening.

Tennessee has been evaluating the possibility of screening since 2006 [8] and we reported our incidence of missed CCHD and the diagnostic gap existing in Tennessee prior to the beginning of state wide screening. [9] Based on our experience and the literature we believed pulse oximetry would be helpful in our state with 80,000 deliveries by detecting 5–

7 babies each year that would otherwise be missed. However, we also calculated that an upper extremity pulse oximetry reading would be unnecessary if an initial foot pulse oximetry reading was 97 or higher since it would be impossible to have a difference of greater than 3%. Therefore, the Tennessee Cardiac Screening Task Force and the Genetics Advisory Committee presented to the Tennessee Commissioner of Health a modified algorithm which had an initial assessment of a single lower extremity reading which if 97 or higher did not require an additional upper extremity second test. This report gives the two year results of this modified screening algorithm.

## Materials and Methods

The Tennessee algorithm starts with a foot only screen, if the pulse oximetry level is 97 or higher the test is a pass and no upper extremity result is obtained. If the level is less than 90 the infant fails and goes to evaluation. Levels of 90–96 then follow the AHA, CDC algorithm. See Figure 2.

The State of Tennessee Health Department has collected data on pulse oximetry screening as part of the State Genetics screening program. The reports are gathered from the pulse oximetry screening form on each blood spot test.

Surveillance for missed cases of CCHD is done through reports from the separate State of Tennessee Birth Defects Registry and the Tennessee Initiative for Perinatal Quality Care (TIPQC) missed CCHD database as previously reported. [9] De-identified information collected by the TIPQC registry includes the neonate's diagnosis, age at diagnosis, presenting symptoms, and outcome. For the purpose of the registry, CCHD was defined as severe and life-threatening CHD requiring either surgical or catheter-based intervention in the first month of life. Reportable lesions include ductal dependent lesions and lesions resulting in hypoxia. Congenital heart defects including acyanotic and non-ductal-dependent congenital heart defects requiring semi-elective surgical repair (Tetralogy of Fallot without cyanosis, atrioventricular septal defect, atrial septal defect, ventricular septal defect, and patent ductus arteriosus) were excluded from the report.

## RESULTS

In 2013 there were 84,964 births and in 2014 an additional 87,184 births for a total of 172,148 births. During that time 95% submitted pulse oximetry screening data (163,699). From that cohort 96% passed with foot only evaluations. This eliminated the need for a second pulse oximetry reading in 156,948 infants.

During the study, 232 infants failed the Tennessee algorithm with 51 or 22% true positives. Eight infants had no prenatal suspicion or clinical signs and were picked up solely by the screen. During the same period there were 13 infants with CCHD who passed the screen. Ten of these had partial left sided obstructive lesions which subsequently presented clinically between day 3 and day 30 of life. Two infants were found to have total anomalous pulmonary venous return (TAPVR) one of whom was found at autopsy. This infant presented at 14 days of life. One infant with no recorded screen presented with cyanosis and Tetralogy of Fallot.

## DISCUSSION

Pulse oximetry screening for CCHD is a valuable adjunct to physical exam and clearly worth the effort. Even in false positive cases a significant number of infants are identified with other disorders that benefit from evaluation and treatment. We did not systematically track all the babies who failed and who did not have CCHD but we found a case of intracranial hemorrhage with apnea, a nasal lacrimal tumor, two babies with pneumonia and one with sepsis. It is clear that no baby should be discharged with a saturation of less than 95 without understanding why. [10]

The Tennessee algorithm saved 150,000 unnecessary pulse oximetry readings and still detected the predicted number of CCHD cases. Using the cost data from Peterson et al [11] and assuming the pulse oximeter probe is not changed between sites would still cost over \$500,000 to obtain an upper extremity pulse oximetry reading which could not be more than 3 % greater than the foot reading. Nursing time is valuable and best used for safe sleep teaching or feeding advice. Besides the cost, job satisfaction is not improved by doing tasks that are futile.

In discussion with physician health care leaders in other states the major objections to using the Tennessee approach is lack of endorsement by the CDC and concern that it is “too complicated”. However, in discussion with nursing leaders it was almost instantaneously understood that if the foot saturation was 97 or higher it was not possible for the hand to be over 3% higher and thus an upper extremity saturation was unnecessary. Thangaratnam et al in their meta-analysis [12] actually showed no population benefit from two extremity testing. We are not recommending eliminating upper extremity testing except in the babies where it could not be helpful. Using two extremity pulsatility differences could help detect aortic coarctations and in the future new technology may allow two site pulse oximetry to detect aortic coarctation by using pulsatility information. [13]

The possibility of a reversal of saturations due to a TGA is not uncommon but this condition would also have to be associated with a saturation of over 97 in the foot. To date there have been no actual reports of such a condition although it is theoretically possible. To require an additional 150,000 pulse oximetry readings each year looking for such a rare condition would not be cost effective. Using cost estimates from Peterson and extrapolating the savings from the Tennessee approach nationwide would result in 3.8 million fewer pulse oximetry tests equating to approximately \$6 million dollars in savings without a loss of screening efficacy. [11]

## CONCLUSION

A staged approach to screening for CCHD is more efficient and will save time and costs. The major drawback to a staged single extremity initial oximetry testing is the need for reeducation and recreation of the multiple, already existent, excellent educational programs. The CDC and AHA policy makers may decide that such effort may not be worth the benefit. However doing 3.8 million unnecessary pulse oximetry tests each year for the foreseeable future may make the reeducation effort worthwhile. Certainly for those states that have not

yet established a pulse oximetry screening program using the most efficient approach would be beneficial.

## Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

## Acknowledgments

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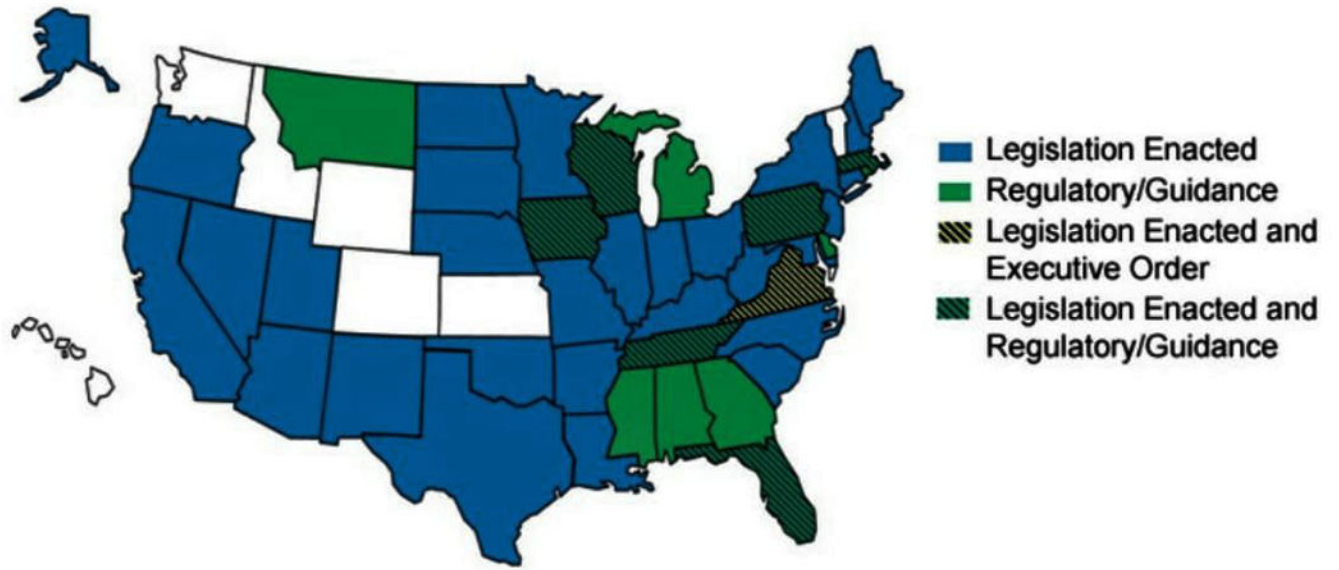
## Abbreviations

<b>AAP</b>	Academy of Pediatrics
<b>AHA</b>	American Heart Association
<b>CHD</b>	Congenital heart disease
<b>CCHD</b>	Critical congenital heart disease
<b>CI</b>	Confidence interval
<b>CoA</b>	Coarctation of the aorta
<b>TIPQC</b>	Tennessee Initiative for Perinatal Quality Care
<b>TN</b>	Tennessee

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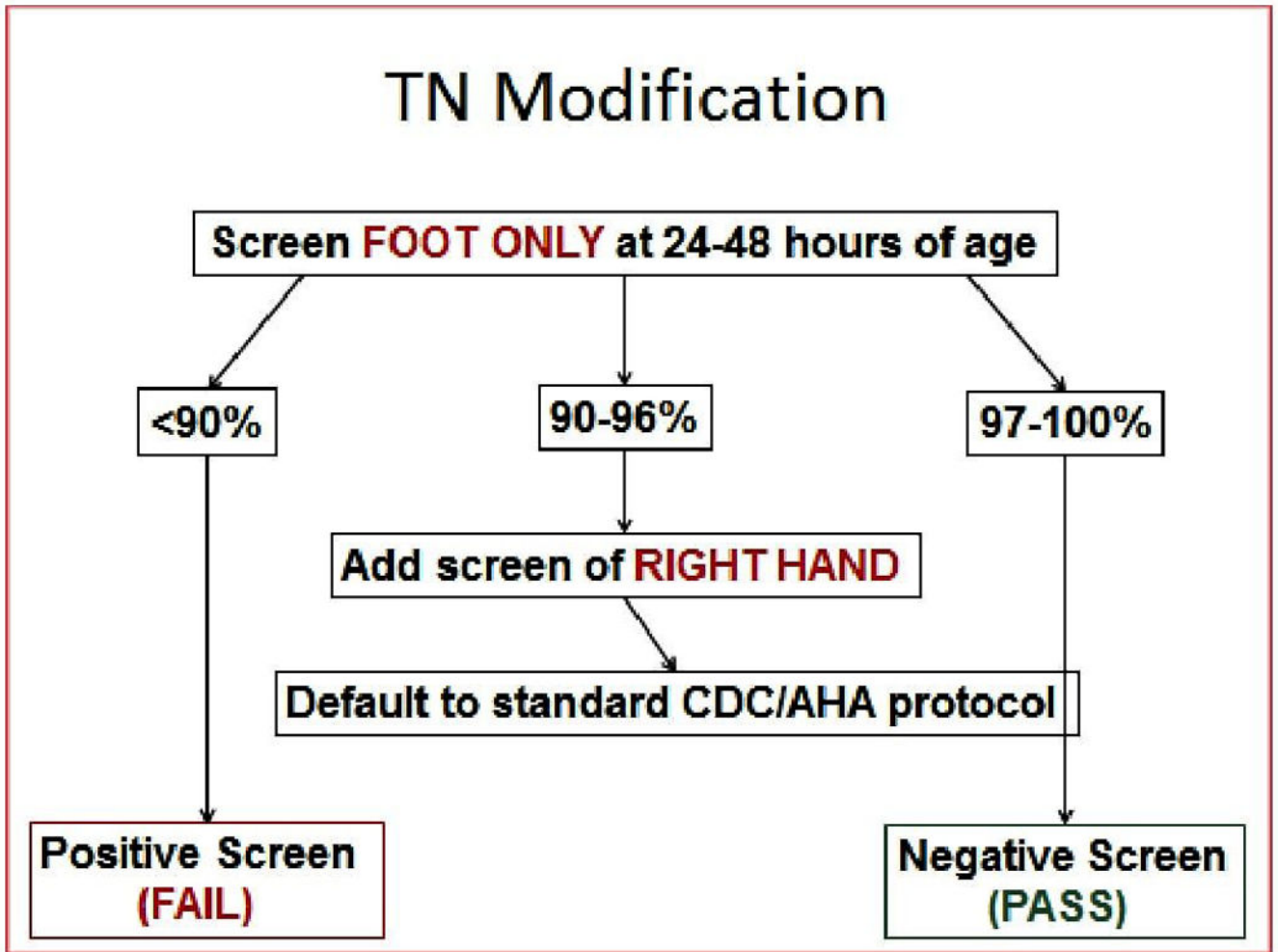
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\*State actions as of December 2014

**Fig 1.**  
State Map of CCHD screening from <http://www.cdc.gov/ncbddd/heartdefects/features/states-adopt-screening.html>



**Fig 2.**  
Tennessee Algorithm. CDC- Center for Disease Control. AHA- American Heart Association