



American Heart Association | **American Stroke Association**

Puget Sound Division
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June 11, 2014

Washington State Board of Health
PO Box 47990
Olympia, WA 98504-7990

Dear State Board of Health Members:

The American Heart Association, in proud partnership with the March of Dimes and the American College of Cardiology, asks for your support to add critical congenital heart disease (CCHD) to Washington's newborn screening panel. We understand forming an advisory group is the first step in the rulemaking process and we respectfully ask you to establish this group to begin this important work without delay.

We understand the Board uses five criteria when considering adding a disorder to the panel; we feel there is strong rationale on all five counts to add CCHD.

- **Public Health Rationale/ Prevention Potential and Medical Rationale:** Congenital heart defects occur in nearly 1 in 100 live births and are the most common cause of infant death, accounting for 27% of infant deaths that are caused by birth defects. In many cases, outwardly healthy infants may be discharged from hospitals before signs of disease are detected. Failure to detect CCHD may lead to serious morbidity or death. These life threatening conditions often require serious medical interventions, in some cases even requiring multiple open-heart surgeries. Evidence suggests that measuring blood oxygen saturation can increase the chances for early diagnosis and detection of CCHD. In 2011, Secretary Sebelius endorsed the addition of CCHD screening to the Recommended Uniform Screening Panel for newborns.
- **Treatment Available:** Once detected, many heart defects can be surgically repaired. It is estimated that 85% of neonates who undergo surgery for CCHD will reach adulthood.
- **Available Technology:** Pulse oximetry detects oxygen levels in the blood. It is a low-cost, non-invasive and painless test - completed in as little as 45 seconds. Pulse oximetry screening for CCHD has a less than one percent chance of giving false positive results, though roughly 25% of infants with low blood oxygen without CCHD may be diagnosed with other conditions that require medical intervention. Pulse oximeters are available in most neonatal units, and hospital staff are trained in how to perform pulse oximetry screening.
- **Cost-Benefit/Effectiveness:** A 2012 cost-effectiveness analysis estimated that universal screening would cost just under \$4 per infant. Research suggests cost savings associated with early detection of a single case of CCHD could exceed the costs associated with screening 2,000 infants. Currently pulse oximetry screening is not billed for but may be included in the bundle of services that hospitals provide.

Adding CCHD to the newborn screening panel is needed to ensure every newborn in Washington is screened. While some may be content with voluntary screening, voluntary compliance today doesn't guarantee screening tomorrow. CCHD is a matter of life and death; this is not the same as the hearing screening. The consequences of missing a baby are too great. Moreover, Washington has high rates of out of hospital births and while data collection is a critical component of a quality screening program, hospital data reporting alone would not guarantee universal screening.

*"Building healthier lives,
free of cardiovascular
diseases and stroke."*

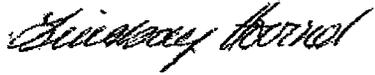
Please remember the American Heart Association in your will.



American Heart Association
June 9, 2014
Page 2

More than 30 other states have established a statewide requirement for screening – including Oregon, Alaska and California. Washington has long been a leader in health care and we urge the Board to begin the rulemaking process to add CCHD to the newborn screening panel and ensure Washington remains a leader in children's health.

Sincerely,



Lindsay Hovind
Government Relations Director

Dear State Board of Health members,

My name is Kelsey Popp, and I'd like to share with you my family's story.

On August 23, 2009, I gave birth to our third beautiful son. My labor and delivery were rushed and a bit stressful, but when my sweet Christian was finally born, I expected to be handed a healthy bouncing baby boy. But instead, Christian was whisked away by our nurses for his newborn screening panel. Christian soon began to cry, and I remember a huge wave of relief, thinking everything was ok. Within moments, the nurse informed me that his initial blood oxygen level (measured by pulse oximetry) was in the low 60's - a normal newborn should be in the upper 90's-100. He would need to go to the nursery for stabilization, and to be transferred from our birth center to the local major hospital's NICU for monitoring. Before his transfer, I was able to sit near his isolette for about an hour. I remember looking at my baby, thinking that on the outside he appeared to be perfectly HEALTHY. There was nothing about him that seemed distressed, and he was pink in color. All I could think was - what if my birth center hadn't measured his blood oxygen level? Would we have noticed anything was wrong before we were discharged to go recover at home?

Christian was transferred to a NICU just a few hours after he was born. Once there, he was given an echocardiogram (an ultrasound of the heart), which diagnosed that our seemingly healthy baby had a very serious heart defect. He had Total Anomalous Pulmonary Venus Return, or TAPVR. His pulmonary veins weren't connected properly, so his heart wasn't pumping oxygen rich blood back to his body. We were informed that without open heart surgery, our precious new son didn't have long to live. When Christian was born, we lived in Hawaii. Unfortunately, Hawaii doesn't have the capacity to do newborn heart surgery. So when Christian was just six days old, he was air-lifted to San Diego. His tiny plane was only big enough to seat a doctor and a couple nurses, so my husband Curtis and I had to fly separately and meet up with our baby in California later that night.

Christian spent the next week and a half battling a fever, and required two blood transfusions during this time. Finally, he was stable enough to undergo open heart surgery when he was sixteen days old. As parents, it was the toughest and scariest day of our lives. But it was also arguably one of the best days; because Christian came through surgery with flying colors, and his defect was able to be fully repaired. On Christian's one month birthday, we flew home to a joyous reunion with our family in Hawaii with a HEALTHY baby!

Today, Christian is almost five years old, and you would never know he had such a rough start at life. He is a fun loving and energetic little boy, who loves to cause chaos right alongside his three brothers every day. He loves cowboys, playing soccer, wearing spiffy bowties, and will be starting kindergarten in the fall. My husband and I are forever grateful that our birth center used pulse oximetry as a regular part of their newborn screening process. We can't bear to think what could have happened if we had been sent home with a baby who appeared healthy, only to have things go south once we were away from a hospital setting. Because of the early pulse oximetry screening and subsequent intervention, Christian was able to get the life-saving care he needed.

Thank you for giving me the opportunity to share Christian's story. As a mother who has seen firsthand how important it is, I urge you to PLEASE make critical congenital heart defect (CCHD) screening a priority. The best, most effective way to initially measure CCHD, is to use pulse oximetry before discharge. It is a quick, painless, non-invasive, and very inexpensive test. If it is made a statewide requirement, pulse oximetry WILL SAVE LIVES of babies here in Washington state.

Sincerely,
Kelsey Popp
788 SW 12th Ct
Oak Harbor, WA 98277



Christian Popp, Age: 4

FACTS

Precious Information

Pulse Oximetry Screening for Critical Congenital Heart Disease

OVERVIEW

Congenital heart defects are malformations of the heart or major blood vessels that occur before birth.¹ In many cases, however, hospital staff may not identify these defects and outwardly healthy infants may be admitted to nurseries and discharged from hospitals before signs of disease are detected.

Congenital heart defects occur in 8 out of 1,000 live births,^{2,3} and are the most common cause of infant death, accounting for 27% of infant deaths that are caused by birth defects.⁴ A quarter of infants who have congenital heart defects will be diagnosed with critical congenital heart disease (CCHD), a life threatening condition that requires surgery or catheter intervention within the first year of life.⁵ Failure to detect CCHD and late detection of CCHD may lead to serious morbidity or death.^{6,7}

Fortunately, an emerging body of evidence suggests that measuring blood oxygen saturation can increase the chances for early diagnosis and detection of CCHD.⁸ Once detected, many heart defects can be surgically repaired. It is estimated that 85% of neonates who undergo surgery for CCHD will reach adulthood.⁹

RECOMMENDED UNIFORM SCREENING PANEL FOR NEWBORNS

Newborn screening is a well-established state-based public health program that involves testing all infants for metabolic, hormonal, genetic, and developmental disorders. Each year, more than 98% of newborns are screened across the United States for these disorders.^{10, 11}

In 2002, the Health Resources and Services Administration (HRSA) commissioned the American College of Medical Genetics to develop a list of conditions that all states could consider including in their screening programs.¹² This list is called the Recommended Uniform Screening Panel¹³ and it currently advises all states to mandate testing for 31 core disorders and 26 secondary disorders. Creation of the Recommended Panel has led to greater uniformity among states in their adoption of screening programs.¹¹ New conditions for screening are frequently nominated for inclusion in the Panel.

In September 2011, the U.S. Secretary of Health and Human Services endorsed the addition of CCHD screening to the Recommended Uniform Screening Panel for newborns.¹⁴ Additionally, the Secretary's Committee on Heritable Disorders in Newborns and Children recommends that hospitals use a specific type of test called pulse oximetry to screen infants for CCHD.¹⁵

CUSTOMARY SCREENING PRACTICE

Several tools are regularly used to identify infants who have heart defects.

- Prenatal ultrasounds performed 18-20 weeks into a pregnancy can reveal anatomical abnormalities.¹⁶ Routine prenatal ultrasounds, however, detect less than 50% of CCHD,⁵ and rates of detection depend on differing levels of access to prenatal ultrasound and degree of practitioner training.⁵
- After birth, infants are physically examined by primary care providers both before hospital discharge and in routine follow-up visits. Physical exam results may lead clinicians to perform additional tests, including chest radiographs, echocardiograms, and pulse oximetry.⁵

Although prenatal ultrasounds and postnatal physical exams successfully detect many heart defects and detection rates have improved since their advent, they are not sufficient to diagnose all cases of CCHD.⁵ New research suggests that when all infants are screened using pulse oximetry in conjunction with the routine practices, CCHD can be detected over in over 90% of newborns.¹⁷

PULSE OXIMETRY SCREENING

Pulse oximetry screening is a low-cost, non-invasive and painless bedside diagnostic test that can be completed by a technician in as little as 45 seconds.⁵ Pulse oximetry testing is conducted to estimate the percentage of hemoglobin in the blood that is saturated with oxygen. When the screening identifies newborns with low blood oxygen concentration, additional testing can be completed to detect heart defects or other life-threatening conditions that could have gone undetected.

Many studies show that pulse oximetry screening for CCHD has a less than one percent chance of giving false positive results.¹⁹ False positive screening results for CCHD can still offer information to doctors: roughly 25% of infants identified as having low blood oxygen without CCHD may be diagnosed with other conditions that require medical intervention.²⁰

The American Heart Association (AHA), the American Academy of Pediatrics (AAP), and the American College of Cardiology Foundation (ACCF) recently outlined recommendations for a standardized pulse oximetry screening approach and diagnostic follow-up.⁸ According to these recommendations, screening should be performed on asymptomatic newborns after 24 hours of life in order to avoid false-positive results.⁸