House Health Care Committee
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February 8th, 2013

Informational Hearing on Critical Congenital Heart Defects/HB 2693

Chair Greenlick, members of the committee, my name is Stephanie Tama-Sweet and I am the Director of Government Relations for the American Heart Association – Oregon. Our mission is to build healthier lives, free of cardiovascular disease and stroke. I am proud to be here today in recognition of Critical Heart Defect Awareness Week and in support of HB 2693.

Congenital Heart Disease (CHD) describes a variety of structural defects that are present at birth. These defects can be asymptomatic (meaning they have no symptoms of disease) or life-threatening. CHDs are the number one birth defect and the number one cause of death in the first year of life.

Oregon does not currently track CHD but extrapolating national statistics with Oregon data from the Oregon Health Authority's Public Health Division has estimated that roughly 300 – 350 babies are born in Oregon each year with CHD. Of these, roughly 75-85 have Critical Congenital Heart Defects (CCHD) which means that the heart defect causes severe, life threatening symptoms and requires intervention within the first hours, days or months of life.

In September of 2011, Kathleen Sebelius, the US Secretary for Health and Human Services endorsed the addition of screening for CCHD with the use of pulse oximetry to the Recommended Uniform Screening Panel (RUSP) for all infants in the US. Pulse oximetry is a non-invasive, painless test that measures the amount of oxygen in the blood. Pulse oximetry is a completely separate test from newborn screening that uses blood spots. It is a bedside test can detect the following 7 CCHD conditions that require intervention:

- Hypoplastic left heart syndrome
- Pulmonary atresia, intact septum
- Tetralogy of Fallot

- Total anomalous pulmonary venous return
- Transportation of the great arteries (vessels)
- Tricuspid atresia
- Truncous arteriosus

Cost estimates for the pulse oximetry test range between $5-10 per infant depending on the equipment used. One recent study estimates that the cost of one missed CCHD case may exceed the cost of screening 2,000 newborns. Many hospitals in Oregon already use pulse oximetry screening for their newborns. Rough estimates from the Oregon Health Authority show about 60% of hospitals are already screening newborns with pulse oximetry tests and many are planning to begin.

Pulse oximetry screening is a simple, cost-effective test that will save lives. Screening all Oregon newborns will help identify heart defects as early as possible, giving these babies the chance to get treatment and prevent future medical complications. Medical costs for one family for their ER visit and NICU stay to get their baby stable enough for surgery can easily total more than $250,000, the vast majority of which could be avoided had the baby been diagnosed in the hospital.

The American Heart Association supports the use of pulse oximetry to detect Critical Congenital Heart Defects and we would like to see every hospital use this screening to give all Oregon newborns the opportunity for a healthy start. There are two bills filed this session, one in the House and one in the Senate that would require the testing of all Oregon babies for CCHD. We strongly these bills and urge your support as well.

Thank you very much and I'm happy to respond to any questions.

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SCREENING FOR CRITICAL CONGENITAL HEART DISEASE

On September 21, 2011, the U.S. Secretary for Health and Human Services (HHS) endorsed the addition of screening for Critical Congenital Heart Disease (CCHD) using pulse oximetry to the Recommended Uniform Screening Panel (RUSP) for all infants in the U.S.[1] How and why was this recommendation made? What does it mean for clinical practice and public health? And where do things stand in Oregon now? Read on, and all will be revealed (or at least what we know and what we don't know).

BACKGROUND

Universal newborn screening involves screening every newborn for certain serious genetic, endocrine, and metabolic conditions (e.g. PKU, sickle cell disease), as well as functional disorders that are not apparent at birth. The goal of newborn screening is to reduce infant morbidity and mortality through early identification and treatment. The Advisory Committee on Heritable Disorders in Newborns and Children (hereafter referred to as "the committee") reviews evidence and provides national guidelines on newborn screening that are reviewed and endorsed by the HHS Secretary. States use the RUSP as guidance when establishing their state-specific screening panels. In 2010, the committee recommended adding CCHD screening with pulse oximetry to the RUSP.[2]

WHY CCHD?

Congenital heart disease (CHD) describes a variety of structural defects that are present at birth. These defects change the normal flow of blood through the heart, and may result in hypoxemia (low blood oxygen saturation) during the neonatal period.[3]

CHD can range in severity from asymptomatic to life-threatening. CHD affects about 7 to 9 of every 1,000 live births in the United States and Europe and is the most common cause of death in the first year of life.[3] (Although we don't have Oregon-specific data, with ~40,000 annual births, this extrapolates to about 300 to 350 CHD cases per year in Oregon). When CHD causes severe and life-threatening symptoms requiring intervention, such as cardiac catheterization or surgery, within the first year of life, it is known as Critical Congenital Heart Disease (CCHD). About one-quarter of neonates with CHD have CCHD[3] (~75-85 cases annually in Oregon. Screening is aimed at identifying and treating newborns with CCHD as early as possible to improve their outcomes.

WHY PULSE OXIMETRY?

Pulse oximetry has several things going for it when it comes to CCHD screening: it's a non-invasive test to estimate hemoglobin oxygen saturation in blood; it's a bedside test; and a positive screen is followed-up by an echocardiogram, just as a physical exam finding would be.[3] It, therefore, has potential to efficiently detect 7 CCHD conditions that require intervention and present most or all of the time with neonatal hypoxemia. These account for about 17-31% of all CHD[3] and were the focus of the committee's review of pulse oximetry screening. They include*:

- Hypoplastic left heart syndrome (HLHS)
- Pulmonary atresia, intact septum
- Tetralogy of Fallot (TOF)
- Total anomalous pulmonary venous return (TAPVR)
- Transposition of the great arteries (TGA)
- Tricuspid atresia
- Truncus arteriosus

EVIDENCE REVIEW

The committee identified 11 studies that addressed the specificity and sensitivity of pulse oximetry screening for CCHD. In all but two, screening was >99% specific (test negative in those without disease) for the seven conditions listed above. Lower specificity (more false positives) appeared to be associated with screening at less than 6 hours after birth and may reflect lower oxygen saturations during the transition to postnatal circulation.[3] Screening was therefore targeted for the second day of life (≥24 hours of age or shortly before discharge if <24 hours of age) in the committee's recommended screening protocol (see Figure 1, *verso)*.

Sensitivity (test positive in those with disease) was more variable, ranging from 42 to 100%. This was thought to be related to differences in the screened populations (e.g. if the study excluded newborns sent to the NICU or newborns who were symptomatic at birth, or if the institution had a large group of prenatal diagnoses) and the testing strategy employed.[3]

RESULTS

The committee determined that pulse oximetry identifies neonates with CCHD that prenatal ultrasound and postnatal clinical assessment miss. One large screening study of close to 40,000 newborns in Sweden found that neonates with CCHD in regions without routine pulse oximetry screening were more likely to be dischaged with undiagnosed ductus arteriosus-dependent circulation (28% vs. 8%) and that neonates diagnosed post-discharge had higher mortality than those diagnosed pre-discharge (18% vs. 0.9%).[3] The committee ultimately recommended that screening combine physical exam and pulse oximetry, as this had the highest sensitivity.

COSTS

Cost estimates for pulse oximetry screening range from less than $5 to $10 per infant, depending on the protocol.[4] This compares favorably with cost estimates for newborn hearing screening, which costs $30 or more per infant.[4] One British study found the cost per timely diagnosis of life-threatening CHD was £4,894 for pulse oximetry.[3] Granted, it's hard to know how that translates to dollars, depending on unit costs for care, exchange rates, or the current state of implosion of the European Union. But it ballparks to around $10,000 in today's dollars, and is likely less costly than com-

* Images available at: www.mayoclinic.com/health/congenital-heart-defects/CC00026 and www.heart.org/HEARTORG/Conditions/CongenitalHeartDefects/AboutCongenitalHeartDefects/Common-Types-of-Heart-Defects_UCM_307017_Article.jsp

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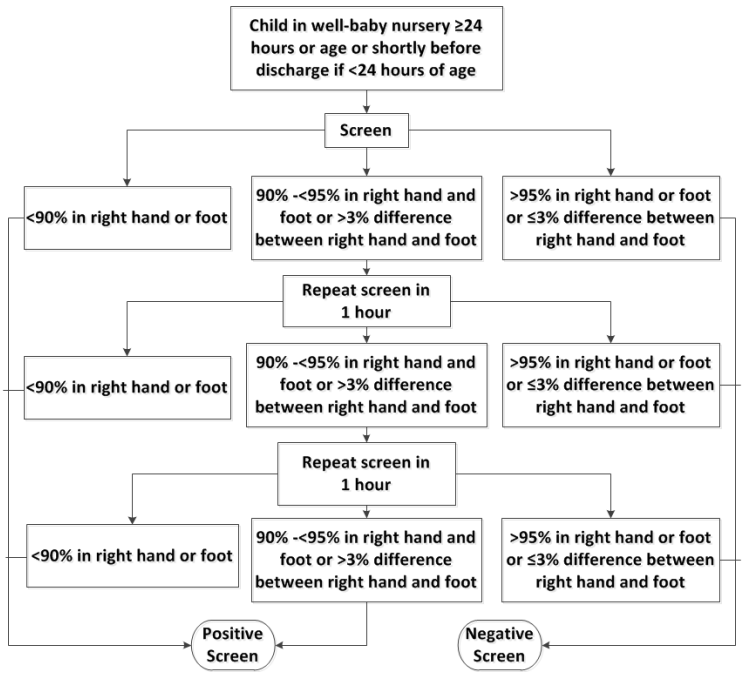
Figure 1. Recommended screening protocol for critical congenital heart disease using pulse oximetry.[4]



plications from undiagnosed CCHD. In fact, the Swedish study mentioned above estimated the cost of complications from one missed CCHD case may exceed the cost of screening 2,000 newborns.[4]

WHAT NOW?

Not to wax too Rumsfeldian on you, but this is where we have some known knowns and some known unknowns. In the known known category was the need for a standardized screening protocol (Figure 1).[4] Known unknowns include: how this protocol may need to be refined as screening data become available or adjusted for special conditions, such as high altitude settings; how to deal with out-of-hospital births; the true benefit of screening when it's done for the general population; the availability of echocardiography or telemedicine services to follow-up positive screening results, particularly in rural areas; and the exact role of public health in quality assurance and surveillance.

We don't have Oregon-specific data on implementation, but anecdotal information from Oregon, Idaho, and SW Washington during 2012 indicates that about 60% of hospitals and birthing centers are currently using pulse oximetry screening, with about 15% of facilities planning to implement it soon.

Despite existing uncertainty, there are a few things we can say for sure:

- Pulse oximetry appears to be both effective and cost-effective for CCHD screening
- Positive pulse oximetry screening should be followed-up with a comprehensive evaluation for causes of hypoxemia[4]
- In the absence of other findings to explain hypoxemia on pulse oximetry screening, CCHD needs to be excluded on the basis of diagnostic echocardiogram; a pediatric cardiologist should be consulted, when feasible before obtaining an echo[4]
- As implementation becomes more complete, primary care providers will need to ensure that all newborns are appropriately screened and receive necessary follow-up.[4]

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