

Critical Congenital Heart Disease Screening & the Heart Smart, Expanding Borders Series

Lisa A. Hom, RN, Esq.

October 2, 2013

Transforming Children's Health Since 1870









Meet our Team









Gerard R. Martin, MD, FAAP, FACC, is a nationally recognized expert in pediatric cardiology. Dr. Martin is a fellow of the American Academy of Pediatrics, the American College of Cardiology and the American Heart Association. In addition, he also served on the American Board of Pediatrics sub-board in pediatric cardiology. Dr. Martin is an associate editor for Pediatric Cardiology, and has more than 100 publications in the field of pediatric cardiology.

Joseph L. Wright, MD, MPH, provides strategic leadership for the organization's advocacy mission, public policy positions and community partnership initiatives. He has served as attending faculty in the Division of Emergency Medicine at Children's since 1993, and recently served the institution as Executive Director of the Center for Hospital-Based Specialties.

Lisa A. Hom, RN Esq. is the Collaborative Practice Facilitator for the Children's National Heart Institute where she oversees, champions and facilitates quality and performance improvement work with clinical teams to improve outcomes for children with congenital heart disease, and for their families. Prior to joining CNHI, she had the unique opportunity to work both as regulatory counsel, focusing primarily on healthcare issues at the state level, and as a pediatric intensive care nurse caring directly for children with critical congenital heart disease.

Lindsay Attaway is the Program Coordinator for the Heart Institute. With a background in graphic design and web development, much of her work is focused on designing materials for the Heart Institute. She will be managing the Congenital Heart Disease Screening Program's online community.





Background

- CCHD screening is expanding rapidly across USA and internationally
- Education identified as an issue
 - What is CCHD screening?
 - How is it performed?
 - Impact of Heart Smart, Expanding Borders videos







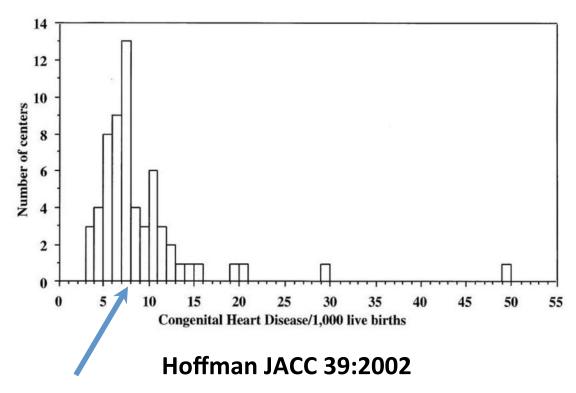






Congenital Heart Disease

- ♥ Most common birth defect
- ♥ 8:1,000 with CHD3:1,000 with CCHD
- ▼ Accounts for ~ 40% deaths fr congenital anomalies
- Majority of deaths due to CH occur in first year of life







Perspective on Importance

Missed Diagnosis of Critical Congenital Heart Disease

Ruey-Kang R. Chang, MD, MPH; Michelle Gurvitz, MD; Sandra Rodriguez, MS

- 15 year retrospective study
- 898 infants died of CCHD in infancy
 152 with missed diagnosis
 299 with late diagnosis
- >50% of CCHD deaths were attributed to late/missed diagnosis
- 30 babies died each year secondary to late diagnosis

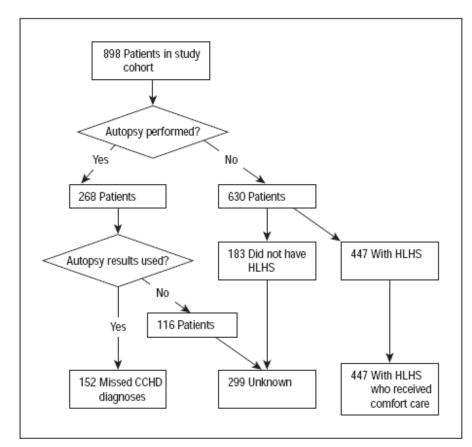


Figure 1. Selection and identification of patients with missed and unknown critical congenital heart disease (CCHD) diagnoses. HLHS indicates hypoplastic left heart syndrome.





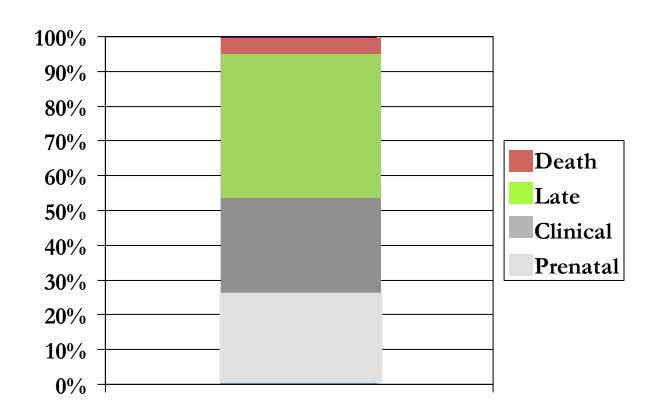
Why is Detecting Newborns with CCHD Difficult?

- Fetal Ultrasound Availability and find rate varies (~23-60%)
- Complex changes from fetal to normal circulation after birth
- Detection through physical examination may be < 50%
 - Auscultation Many FP and FN
 - Palpation of Pulses Depends on patency of PDA
 - Cyanosis Difficult for HCP to detect





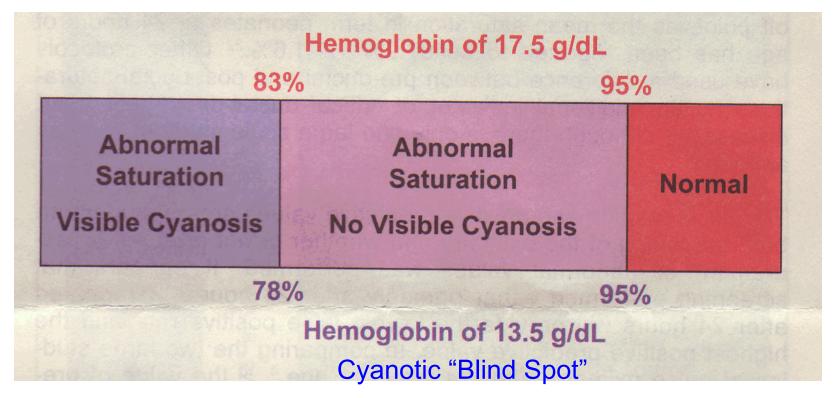
Diagnostic Gap







Difficulty Detecting Cyanosis



Mean threshold for detection 69%!





Pulse Oximetry as a Screening Method

- ♥ Pulse oximetry measures oxygen saturation of hemoglobin in arterial blood
- ♥ Non-invasive and painless test

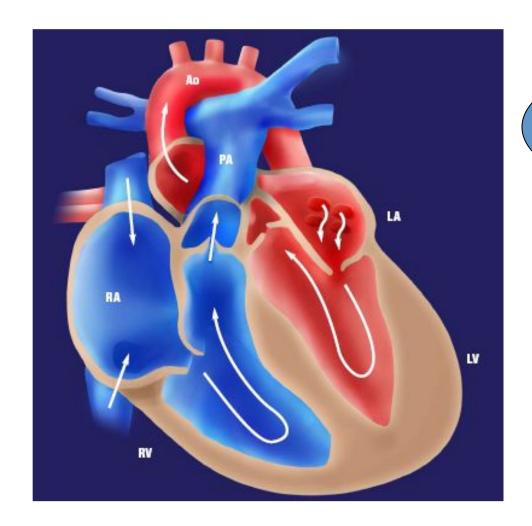


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Normal Newborn Circulation



Passing Sat 100%





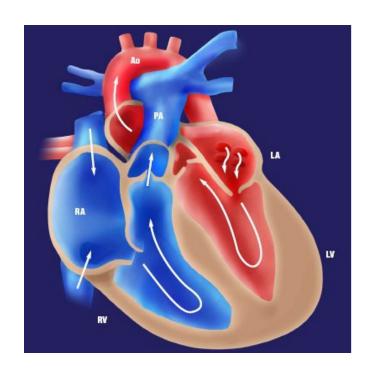
CCHD Screening Primary Targets

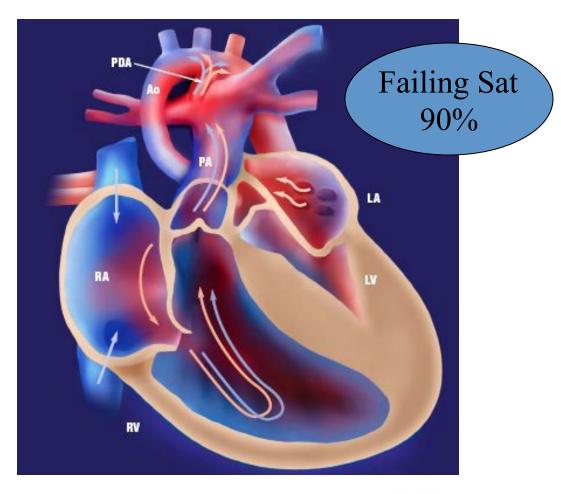
- 1. Hypoplastic Left Heart Syndrome
- 2. Pulmonary Atresia (with intact septum)
- 3. Tetralogy of Fallot
- 4. Total Anomalous Pulmonary Venous Return
- 5. Transposition of the Great Arteries
- 6. Tricuspid Atresia
- 7. Truncus Arteriosus





Hypoplastic Left Heart Syndrome









Secondary Target: Pneumonia







2009 Scientific Statement

- Critical CHD is not detected in some infants
- Failure to detect is associated with significant morbidity and occasional mortality
- Pulse oximetry may detect critical CHD

American Heart Association and American Academy of Pediatrics





BMJ

Impact of pulse oximetry screening on the detection of duct dependent congenital heart disease: a Swedish prospective screening study in 39 821 newborns

Table 2|The performance of screening methods in the detection of duct dependent circulation in newborn infants in West Götaland (1 July 2004 to 31 March 2007)

Performance	Physical examination alone (n=38374)	Pulse oximetry (n=38429)	Physical examination plus pulse oximetry (n=38429)
Sensitivity (95% CI) (%)	62.50 (35.43 to 84.80)*	62.07 (42.3 to 79.31)	82.76 (64.23 to 94.15)
Specificity (95% CI) (%)	98.07 (97.93 to 98.21)	99.82 (99.77 to 99.86)	97.88 (97.73 to 98.03)
Positive predictive value (95 % CI) (%)	1.35 (0.65 to 2.47)	20.69 (12.75 to 30.71)	2.92 (1.88 to 4.31)
Negative predictive value (95% CI) (%)	99.98 (99.96 to 99.99)	99.97 (99.95 to 99.99)	99.99 (99.97 to 100.00)

Table 3 | Pathology found in 69 babies with false positive results from pulse oximetry screening for duct dependent circulation in West Götaland (1 July 2004 to 31 March 2007)

		Subsequent management			
	No (%) of babies	Stay in neonatal intensive care			
Pathology found		≥5 days after screening	<5 after screening	Follow-up only	Surgery
ther critical congenital heart disease*	4 (6)	4/4	0/4	0/4	4/4
Other milder congenital heart disease	10 (14)	4/10	1/10	5/10	4/10
Persistent pulmonary hypertension	6 (9)	3/6	0/6	3/6	N/A
ransitional circulation†	8 (12)	0/8	3/8	2/8	N/A
nfections	10 (14)	6/10	4/10	N/A	N/A
Pulmonary pathology	7 (10)	5/7	1/7	1/7	N/A
Normal (verified from hospital charts)	24 (35)	N/A	N/A	N/A	N/A

*Aulmonary atresia with multiple aorto-gulmonary collaterals (n=2), tricuspid atresia with pulmonary stenosis and ventricular septal defect (n=1), total anomalous pulmonary venous return (n=1).

†Right to left shunting across feramen ovale without pulmonary hypertension.

Granelli BMJ 338:2009





Is Pulse Oximetry Effective in Detecting CCHD?

Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis

Shakila Thanaaratinam Kiritrea Rrown Javier Jamara Khalid SKhan Andrew K Fwer

-13 primary studies - 229,421 infants

screened

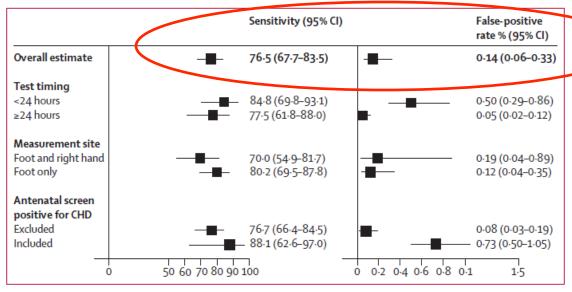


Figure 3: Accuracy estimates based on clinical and test characteristics of pulse oximetry in detection of critical congenital heart defects in newborn babies CHD=congenital heart defects.

Interpretation Pulse oximetry is highly specific for detection of critical congenital heart defects with moderate sensitivity, that meets criteria for universal screening.





CCHD Screening Feasibility in Community Hospitals

Open

Journal of Perinatology (2012), 1–6 © 2012 Nature America, Inc. All rights reserved. 0743-8346/12



- No Additional Staff Required

- Avg. Time to Screen 3.5 minutes (0-35 min)

- 98% of eligible were screened (6,745/6,841)
- Barriers reported in 1.4% (97/6,745)

ORIGINAL ARTICLE

Feasibility of implementing pulse oximetry screening for congenital heart disease in a community hospital

EA Bradshaw¹, S Cuzzi^{1,2,3}, SC Kiernan², N Nagel², JA Becker^{1,3} and GR Martin^{1,3}

¹Children's National Medical Center, Washington, DC, USA; ²Holy Cross Hospital, Silver Spring, MD, USA and ³The George Washington University School of Medicine, Washington, DC, USA

Objective: Pulse oximetry has been recognized as a promising screening tool for critical congenital heart disease (CCHD). The aim of this research was to study the feasibility of implementation in a community hospital setting.

Study Design: Meetings were conducted to determine an implementation plan. Pulse oximetry was performed on the right hand and foot after 24 h of age. Newborns with a saturation $\leq 95\%$ or a $\geq 3\%$ difference were considered to have a positive screen. Screening barriers, screening time and ability to effectively screen all eligible newborns were noted.

Result: From January 2009 through May 2010, of 6841 eligible newborns, 6745 newborns (98.6%) were screened. Of the nine infants with positive pulse oximetry screens, one had CCHD, four had CHD and four others were determined to have false positive screens. Average screening time was 3.5 min (0 to 35 min).

Conclusion: Pulse oximetry can be implemented successfully in

care have resulted in a significant reduction in mortality secondary to CCHD; however, timely diagnosis remains an issue for these newborns. Despite prenatal diagnosis and newborn examinations, as many as 39% of infants diagnosed with CCHD are diagnosed only after discharge from the newborn nursery. Delay in diagnosis may have significant adverse implications; one study showed that 43% of cases diagnosed after hospital discharge from the nursery were in shock at the time of readmission.

Pulse oximetry has been recommended as a potential newborn screening test for CCHD. Early efforts provided the conceptual basis for pulse oximetry in the detection of CCHD.⁵⁻⁹ Subsequent work has provided additional evaluation of the sensitivity, specificity and diagnostic gap of pulse oximetry screening. ^{10–14} In 2009, the American Heart Association (AHA) and American Academy of Pediatrics (AAP) released a statement on the potential use of pulse oximetry screening to detect CCHD. ¹⁵ The statement recognized that the most favorable outcomes are realized when screening on the right lower extremity is conducted after





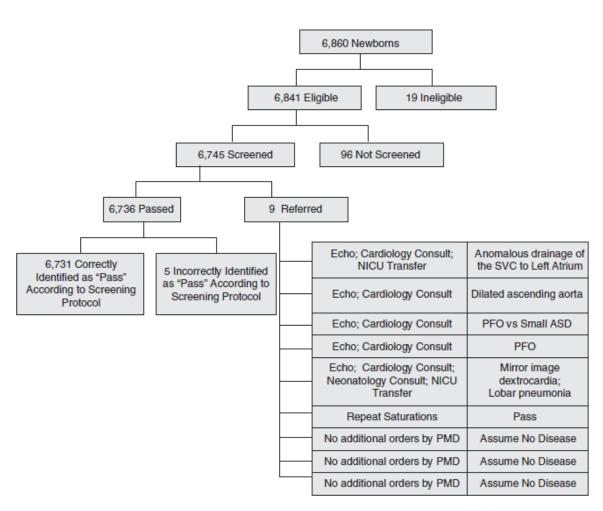
CCHD Screening Feasibility in Community Hospitals

- Avg. Pox Sat

Rt Hand/Foot 100% (90-100%)

Difference 0.2% (0-6%)

-CCHD screening did not lead to a significant increase in echos







Parent Perspective?

Pulse oximetry screening for congenital heart defects in newborn infants: an evaluation of acceptability to mothers

Rachael Powell,¹ Helen M Pattison,¹ Abhay Bhoyar,² Alexandra T Furmston,³ Lee J Middleton,³ Jane P Daniels,³ Andrew K Ewer^{4,5}

Acceptable to mothers FP did not increase anxiety

Design and setting A questionnaire was completed by a cross-sectional sample of mothers whose babies were recruited into the PulseOx Study which investigated the test accuracy of pulse oximetry screening.

Participants A total of 119 mothers of babies with false-positive (FP) results, 15 with true-positive and 679 with true-negative results following screening.

Main outcome measures Questionnaires included measures of satisfaction with screening, anxiety, depression and perceptions of test results.

Results Participants were predominantly satisfied with screening. The anxiety of mothers given FP results was not significantly higher than that of mothers given true-negative results (median score 32.7 vs 30.0, p=0.09). White British/Irish mothers were more likely to participate in screening, with a decline rate of 5%; other ethnic groups were more likely to decline with the largest increase in declining being for Black African mothers (21%, OR 4.6, 95% CI 3.8 to 5.5). White British mothers were also less anxious (p<0.001) and more satisfied (p<0.001) than those of other ethnicities **Conclusions** Pulse oximetry screening was

acceptable to mothers and FP results were not found to increase anxiety. Factors leading to differences in participation and satisfaction across ethnic groups need to be identified so that staff can support parents appropriately.





United States Efforts

- ♥ Health Resources and Service Administration's Secretary's Advisory Committee on Heritable Disorders in Newborns and Children; October 2010
- ♥ HRSA Workgroup Meeting on Screening at ACC Heart House; January, 2011









THE SECRETARY OF HEALTH AND HUMAN SERVICES WASHINGTON, D.C. 20201

September 21, 2011

♥ CCHD Screening endorsed by Secretary Kathleen Sebelius; September, 2011

- ♥ AHA, ACC, AAP and March of Dimes all endorse CCHD Screening
- ♥ 2nd Stakeholders Meeting February, 2012. Publication with outcomes accepted by Pediatrics.

R. Rodney Howell, M.D. Committee Chairperson Secretary's Advisory Committee on Heritable Disorders in Newborns and Children 5600 Fishers Lane, Room 18A19 Rockville, MD 20857

Dear Dr. Howell:

As indicated in my letter to you on April 20, 2011, I determined that the Secretary's Advisory Committee on Heritable Disorders in Newborns and Children's (SACHDNC) recommendations pertaining to the addition of Critical Congenital Heart Disease (CCHD) screening to the Recommended Uniform Screening Panel (RUSP) were not yet ready for adoption.

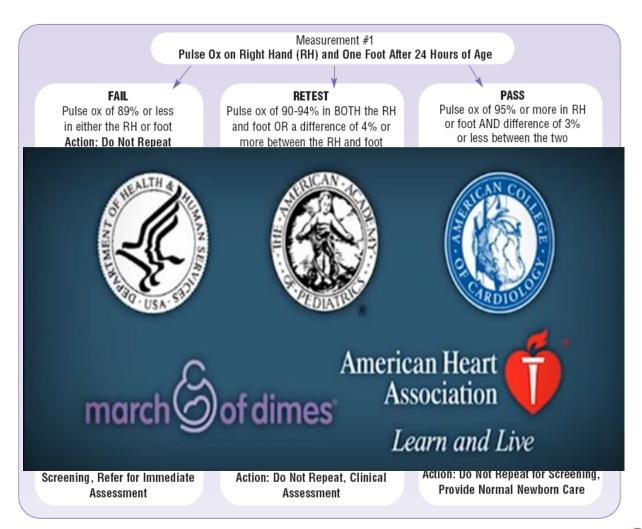
Consequently, I referred the SACHDNC's recommendations to the Interagency Coordinating Committee on Screening in Newborns and Children (ICC) for additional review and input regarding implementation. I asked the ICC to review the evidence gaps described by the SACHDNC and propose a plan of action to address: identification of effective screening technologies, development of diagnostic processes and protocols, education of providers and the public, and strengthening service infrastructure needs for follow-up and surveillance. I have received and reviewed the requested ICC Plan of Action.

As you know, congenital heart disease causes up to 3% of all infant deaths in the first year of life. Heart defects affect about 7 to 9 of every 1000 live births, one quarter of which could be detected and potentially treated by measuring blood oxygen saturation. Given this reality and the available information on the effectiveness of screening, I have decided to adopt the SACHDNC's first recommendation to add CCHD to the RUSP. In addition, I am requesting that the SACHDNC collaborate with the Health Resources and Services Administration (HRSA) to complete a thorough reclusion of the potential public health impact of universal screening for CCHD, as required by the authorizing statute, section 1111 of the Public Health Service Act (42 U.S.C. § 300b-10(b)(4)).





United States Efforts







United States Efforts



- 2011: Indiana and Maryland first states to pass CCHD screening legislation. New Jersey first state to implement universal CCHD screening.
- 20 states enacted legislation in the first half of 2013
- 33 states total with legislation pending in many more

Maryland Bill Signing May 19, 2011





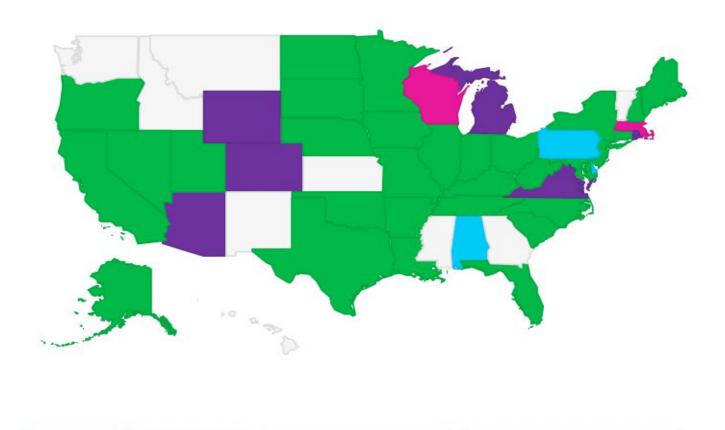
United States Efforts June 2012

cchdscreeningmap.com newborn coalition Washington Montana : Legislation Passed New Jersey : Legislation Introduced : Legislation Pending : Legislation Introduced & Multi-Center Screening and/or Pilots : Legislation Passed & Multi-Center Screening and/or Pilots : Multi-Center Screening and/or Pilots : No Action States 42%





United States Efforts Sept 2013





Legislation Enacted

Regulatory Addition to NBS Panel

Multi-Center Screening or Pilot Project





Congenital Heart Disease Screening Program: Health Authority of Abu Dhabi

New screening saves 13 babies in Abu Dhabi

Implementation Began:

January, 2011

Infants Screened: Approx. 23,000

near 50,000

Total Detected:

13 with CCHD Detected

now 21 detected

SIMPLE TEST IS NOW MANDATORY IN ALL HOSPITALS IN THE CAPITAL

ABU DHABI

By Saminan Zaman Staff Reporter

critical shortage of paediatric cardiologists in the emirate of Abu Dhabi is putting the lives of hundreds of babies at risk, especially if a mandatory test to detect serious congenital heart abnormalities is not administered, health experts have said.

The test, which screens for critical congenital heart disease, affecting three in every 1,000 babies worldwide, is important because babies in the GCC could present certain abnormalities that can kill them within the first two months



Abdul Rahman/Gulf News

Happy and healthy

 Dr Gerard Martin of the Children's National Medical Centre, with 8-month-old Jumana and her father Surour Khamis Abdullah.





International Partners





MOH Hospitals Kuwait



Ahmadi Hospital Kuwait
Oil Company



Hamad Medical Corporation



Health Authority of Abu Dhabi (HAAD) (23 Birthing Hospitals)



Saudi Arabia

Philippines, India, Australia, British Columbia, Great Britain, Mexico





European Efforts: Strategizing for a Uniform Recommendation

THE LANCET

Pulse oximetry screening for congenital heart defects

The Lancet, Volume 382, Issue 9895, Pages 856 - 857, 7 September 2013

*Andrew K Ewer, Anne De-Wahl Granelli, Paolo Manzoni, Manuel Sánchez Luna, Gerard R Martin a.k.ewer@bham.ac.uk

Germany

Italy

Netherlands

Spain

Sweden

UK









What's Next?

HRSA Demonstration Projects June, 2012-2015
 NH, UT, WI, MI)

(VA, NJ,

- Health Information Exchange to facilitate accurate and compatible transfer of data from oximetry device to end-user (Hospitals, DOH, Medical Home, Families)
- "Smart" Oximetry devices to guide screening and provide QA measures
- Additional research efforts CDC, NIH

Widespread implementation and need for education both nationally and internationally...





Heart Smart, Expanding Borders

Year 1 Goals for 2012:

- Web based videos
 - Provider video
 - Parent and screening advocate video

Year 2 Goals for 2013:

- Improve access
 - Creating DVDs of each of the videos available at no charge
 - Large and growing number of non-English speaking families
 - International pulse oximetry screening community

Overall Goal: Free educational tool with widespread access resulting in increased detection of CCHD through the screening of infants prior to leaving the newborn nursery.







Heart Smart, Expanding Borders

- Provider Video content:
 - How & why screening is performed
 - Physiological changes
 - Potential complications
 - How to educate parents including resources and key points of emphasis





Screening in Your Unit

- Pair screening with other standard-of-care newborn screening
- Conduct screening in quiet area and, if possible, with parent present to soothe and comfort the infant.
- Screen while infant is awake and quiet.
- Do not attempt to perform pulse oximetry on an infant while he or she is crying or cold.
- Know the "Dos" and "Don'ts" of Pulse Oximetry







Screening in Your Unit: Pulse Ox Dos



- 1. New, clean sensor for each infant
- 2.Best sites for infants are great toe, thumb and outer aspect of palm and the foot
- 3. Sides of sensor should be directly opposite each other
- 4.Skin should be clean and dry (skin color and jaundice do not affect the pulse ox reading)
- 5. Swaddle and encourage family involvement, if possible make sure the infant is awake.
- 6. Use the confidence indicators to ensure reading is accurate





Screening in Your Unit – Pulse Ox Don'ts

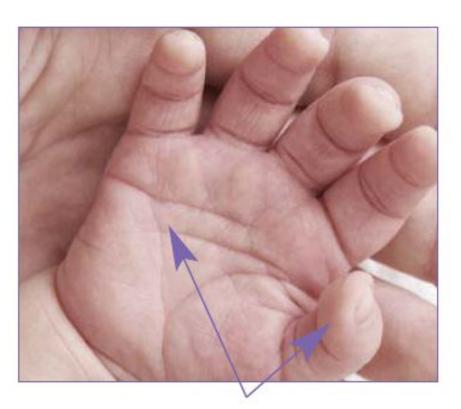


- 1. Never use an adult pulse ox clip for an infant
- 2. Do not use limb with Blood Pressure cuff
- 3. Do not perform in bright or infrared light (may need to cover sensor with a blanket to ensure accuracy of reading)
- 4. No not use tape or your own hand to apply the pulse ox sensor to the infant's skin.





Screening in Your Unit



RH Application Sites



Foot Application Sites





Screening in Your Unit

Application with Disposable Probe



© Masimo Corporation 2011

Application with Reusable Probe







Heart Smart, Expanding Borders

Parent Video content:

- -Family story and interviews
- -How & why screening is performed
- -What are next steps if screen is positive





Parent Education

- Inform the parent or guardian that the purpose of the screening program is to screen for serious heart problems in babies.
- Inform the parent or guardian that the baby will be screened after he or she is 24 hours old.
- Inform the parent or guardian that if she agrees to have the baby screened the pulse ox test will be done on the baby's right hand and one foot, if possible.
- Inform the parent or guardian that the pulse ox test is not usually painful and that it only takes a few minutes when the baby is quiet, warm and not moving.
- Inform the parent or guardian that it is possible that a baby with a heart problem may have a normal pulse ox reading.

- Inform the parent or guardian that they have the right to decline screening.
- Inform the parent or guardian that they may ask questions at any time before, during, or following the screening.







Heart Smart, Expanding Borders

SIGNS OF CHD MAY INCLUDE:

- Tires easily or falls asleep during feeding
- Sweating around the head, especially during feeding
- Fast breathing when at rest or sleeping
- Pale or bluish skin color





Signs of CHD continued

- Poor weight gain
- Sleeps a lot or not playful or curious for any length of time
- Puffy face, hands, and/or feet
- Often irritable or difficult to console





Baby's First Test









Heart Smart, Expanding Borders 2013

Table 1. Detailed Languages Spoken at Home and Ability to Speak English for the Population 5 Years and Over for the United States: 2006-2008 Release Date: April, 2010

			Spoke English		
	Number of	Margin of	less than	Margin of	
	speakers	Error ¹	"Very Well"	Error ¹	
Population 5 years and over	280,564,877	7,708	24,252,429	67,280	
Spoke only English at home	225,488,799	83,368	(X)	(X)	
Spoke a language other than English at home	55,076,078	81,124	24,252,429	67,280	
Spoke a language other than English at home	55,076,078	81,124	24,252,429	67,280	
		 			

- Culturally sensitive
- Need to create solutions for underserved populations





Newly Translated Parent Versions

The Videos:

Pulse Ox for Providers (English only)
Pulse Ox for Parents (English)



Heart Smart: CCHD Screening for Parents in:

- Arabic
- Chinese
- French
- Russian
- Spanish

Available online at:

http://www.youtube.com http://www.babysfirsttest.com http://vimeo.com







Educational Opportunities

Broad stakeholder dissemination:

 State Departments of Health, Nursing Schools, Parent advocacy groups, hospitals, professional organizations

Providers

- Grand Rounds, conferences
- Skills days, orientation

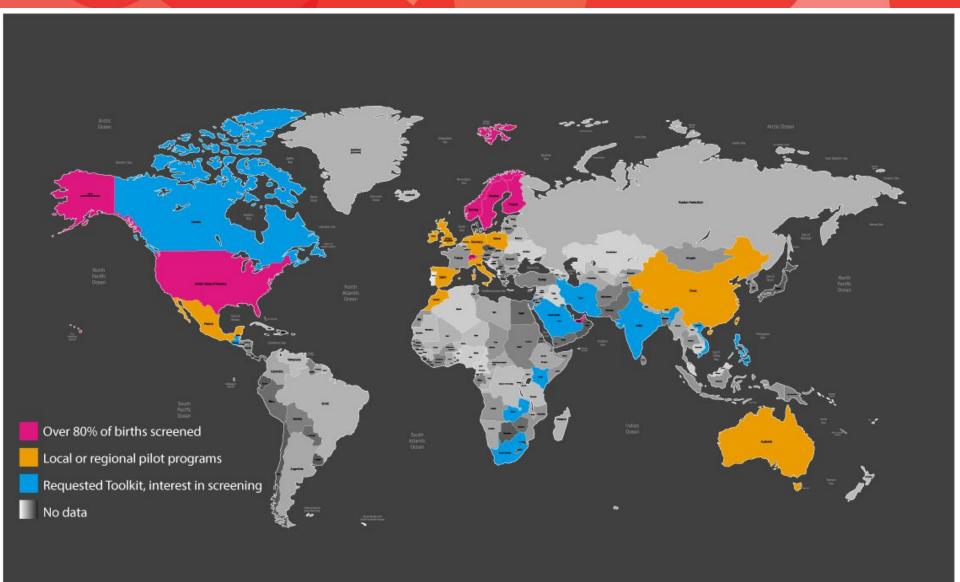
Parents

- Pre-natal tours & birthing classes
- New parent classes on post-partum units













How do I get copies?

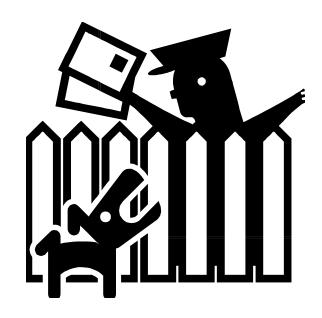
To request hard copy DVDs of the Heart Smart, Expanding Borders education videos send an email to: pulseox@cnmc.org

Please include:

- Your name
- Your organization
- Your complete address

Number of copies & specify type of videos

- Parent or Provider?
- Which languages?





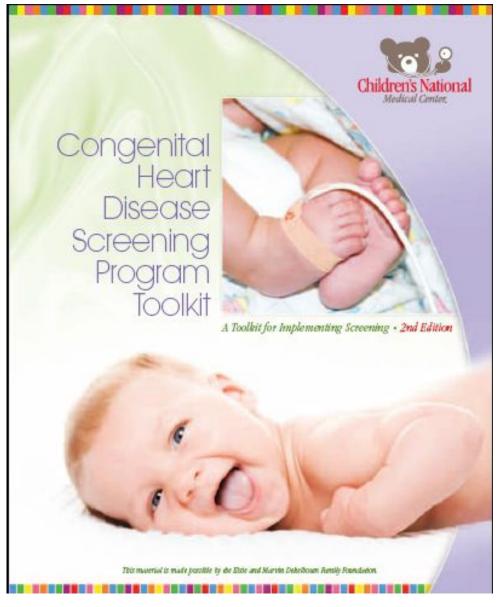


Vision

All infants with critical congenital heart disease are detected before leaving the newborn nursery.

Toolkit Includes:

- Implementation Recommendations
- Screening Protocol
- Education for Families
- Competencies for Providers
- Advocacy Resources and Stories







Online Community Resources

Pulse Ox section under the CNMC's website and our online community dedicated to CCHD Screening.





www.childrenshealthcommunities.org/pulse_ox





Other Resources

Center for Disease Control

www.cdc.gov/ncbddd/pediatricgenetics/cchdscreening.Html



http://www.babysfirsttest.com/

Parent Advocacy Groups

http://1in100.org/; www.tchin.org

Newborn Coalition

http://newborncoalition.com/

ACMG ACTion Sheet











Contact Information

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With thanks to Elizabeth Bradshaw Mikula!



