Am I Blue?
Newborn Pulse Oximetry Screening for Critical Congenital Heart Disease
"The State of the State"

Brian P. Youth, MD
Kelley Bowden, MS, RN

The Barbara Bush Children's Hospital at MMC
Maine AAP Spring Conference
May 5th, 2013

The Barbara Bush Children's Hospital
At Maine Medical Center

Overview

- Briefly review the data to support screening
- Maine's approach to screening
- Regional Collaboration
- Data Review to date
- What's working, what's not
- Legislative Initiative and Issues

Objectives

- State rationale for screening newborns for Critical Congenital Heart Disease
- Describe screening process
- Discuss materials available to support implementation of Critical Congenital Heart Disease screening at your facility

Points to leave with

- Newborn saturation screening is happening in ME
- Clinical examination misses critical CHD
- Oximetry is stable and reliable
- False positive rates are low (lower than false positive rate based on physical exam)
- Data to support this is strong
- Some lesions will not be detected

Overview

- CHD leading cause of infant death
  - 40% of all deaths from congenital defects
  - 3-7.5% of infants deaths are due to cardiac anomalies
- Failure to detect early increases the risk of circulatory collapse
  - Adverse effect on prognosis
  - Poor clinical status at time of surgery increases surgical mortality

Congenital Heart Diseases: The magnitude of the problem

CHD:

5-10 / 1,000 live births

1.4 cyanotic CHD / 1,000 live births

2 critical CHD / 1,000 live births

25,000 cases of CHD/yr in US

25% of infantile deaths

31% of neonatal deaths

Disease	Incidence
Hemoglobinopathy	1/2,000
Cystic fibrosis	1/2,000
Hypothyroidism	1/4,000
Phenylketonuria	1/10,000
Adrenal hyperplasia	1/12,000
MCAD deficiency	1/17,000
Galactosemia	1/50,000
Biotidinase deficiency	1/100,000
Tyrosinemia	1/100,000
Homocystinuria	1/150,000
Maple syrup urine disease	1/180,000
Miscellaneous amino acid, fatty acid, organic acid, and lysosomal abnormalities	1/50,000 to <1/100,000
All together	1.55 /1,000 live births

Heron, M., et al. (2009). Deaths: Final data for 2006. National Vital Statistics Reports, 57(14). U.S. CDC and Prevention.

http://www.cdc.gov/ncbddd/features/heartdefects-keyfindings2010.html

Screening strategies for Critical Heart Disease

- Physical examination
- Fetal echocardiography
- Oximetry
  - Most critical heart disease produces some degree of cyanosis not visible to examiner
  - To date uncertainty exists regarding false positive rate and test accuracy
  - Recent meta-analysis in Lancet May 2012 provides important data in 229,421 newborn babies

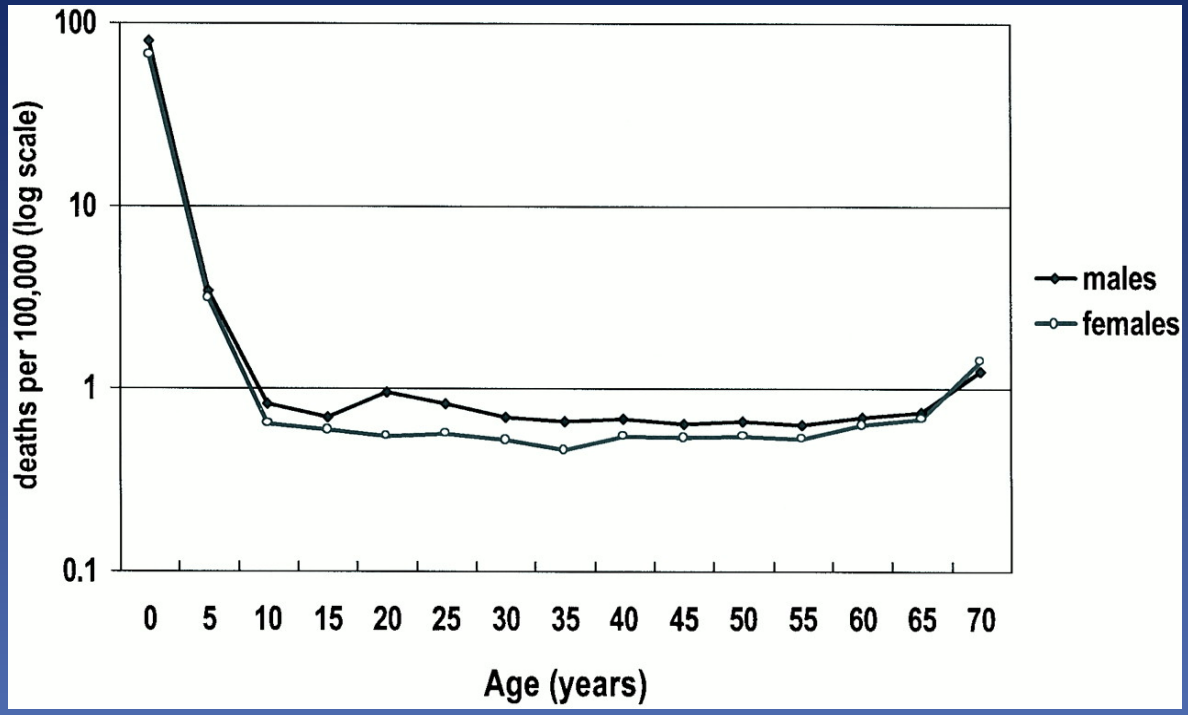
- Critical heart disease
  - Duct dependent systemic or pulmonary circulation
  - Surgery required within 28 days of birth
  - Examples
    - HLHS, PA-IVS
    - Aortic Stenosis
    - TAPVR
    - Severe coarctation

Missed Critical Congenital Heart Diseases (CCHD)

First author	Years	CCHD				Missed CCHD		
		live born		/1,000 live births	deaths	n	% CCHD	% deaths
		prenatal Dx	postnatal Dx					
Aamir [19]	1999–2004	18	94	0.2	–	47	50	–
Abu-Harb [22]	1985–1990	–	–	–	185	56	–	30
Brown [4]	1999–2002	56	230[1]	–	–	73	32	–
Chang [21]	1989–2004	–	–	–	898	152	–	17
de Wahl-Granelli [6]	2004–2007	2	60[2, 3]	1.3		19	32	
		9	109[4]	1.0		28	28	
Koppel [16]	1989–1999	9	11	1.8		3	27	
Kuehl [20]	1981–1989	–	4,390	–	800	76		9.5
Liske [17]	2000–2002	–	62[5]	2.78	–	15	25	–
			110[6]	–		–	–	
Meberg [23]	2005–2006	31	50[7]	1.2	–	6	12	–
		7	48[8]			11	23	
Mellander [24]	1993–2001	–	259[9]	–	–	51	20	–
Schultz [25]	2000–2003	31	45[10]	–	–	12	27	–
Wren [15]	1985–2004	55	614	0.97	–	198	32	15[11]

Hoffman, J. It is time for routine neonatal screening by pulse oximetry. Neonatology 2011;99:1-9

Timing of death from CHD



50% of deaths from CHD occur in 1st year and

50% of infantile deaths occur in 1st month of life

Boneva, R: Circulation. 2001;103:2376

Timeline of pulse oximetry screen for CCHD

Large European prospective studies
(Norway, Sweden, UK, Germany, Switzerland)

oximetry screen
1st clinical reports

Larger prospective study
In NY (Koppel)

Several European countries
adopt pulse-ox screen
as standard of care

AAP endorses HHS
recommendations

Request to AHA
For recommendations

SACHDNC (HHS)
recommended
adding pulse-ox screen

AHA comment
Evidence not sufficient

Maine Hospitals start
screening

AHA/AAP
statement comment

Proposed bill in TN
to mandate screening

2002 **2005** **2008** **2011** **2012**

Workgroup convened…

- Primary Care Providers, Pediatric Cardiologists and neonatologists, nurses, AAP, ACCF, AHA, ACMG, March of Dimes, Assoc. of Maternal and Child Health Programs, Association of Public Health Laboratories, the SACHDNC, parent screening advocates, state public health officials, CDC Reps, USFDA, HRSA, and NIH
- Led by William Mahle, MD* and R. Rodney Howell, MD**
  - *Led the development of the AAP/AHA statement
  - **Chair of the SACHDNC
- Meeting focused on recommendations for pulse-oximetry monitoring for CCHD including recommendations for the service infrastructure needs for f/u, and strategies for education

SACHDNC recommendations

- ♥ Research:
  - ♥ NIH shall fund research activities to determine the relationships among the screening technology, diagnostic processes, care provided, and then health outcomes of affected newborns with CCHD as a result of prospective newborn screening
- ♥ Surveillance:
  - ♥ CDC shall fund surveillance activities to monitor the CCHD link to infant mortality and other health outcomes

SACHDNC recommendations

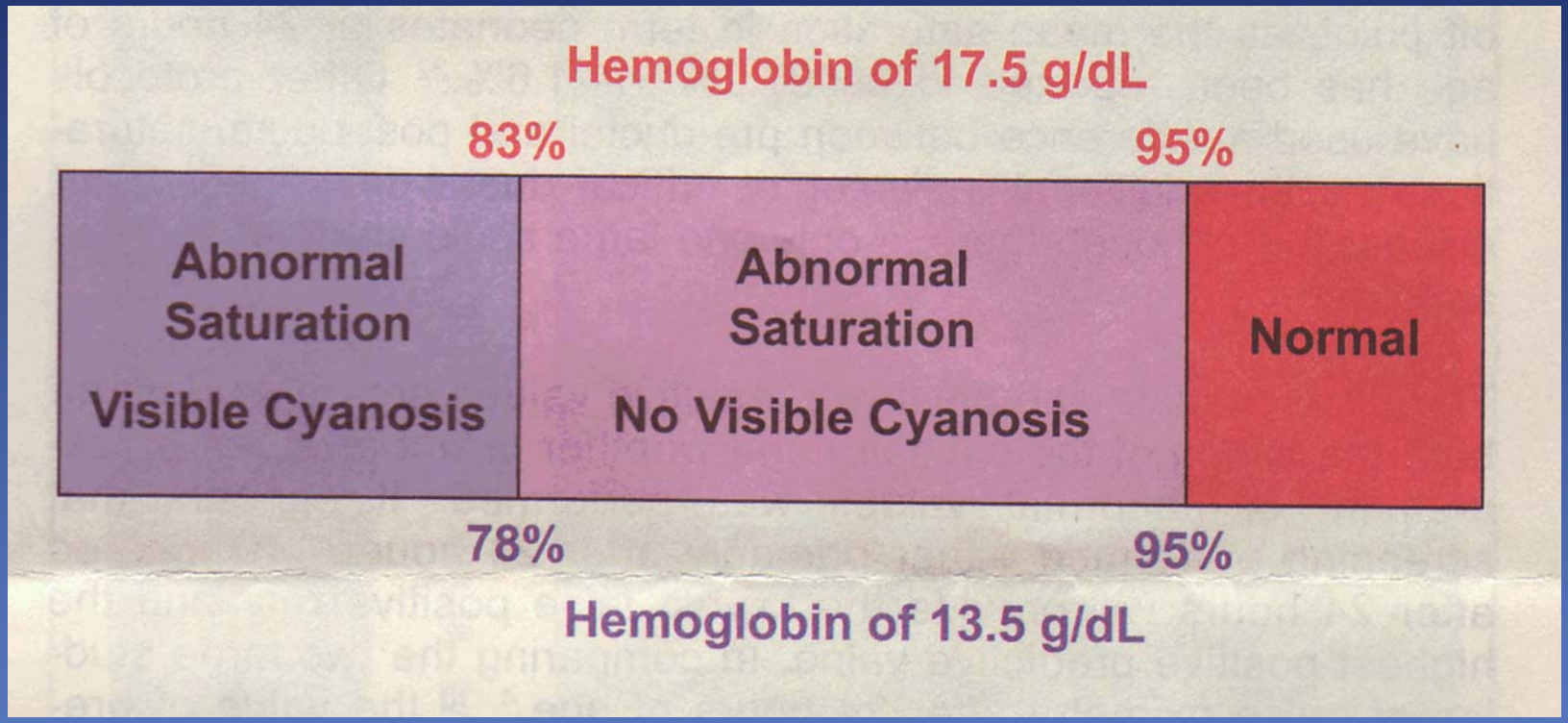
- ♥ Screening Standards and Infrastructure:
  - ♥ HRSA shall guide the development of screening standards and infrastructure needed for the implementation of a public health approach to point of service screening for Critical Congenital Cyanotic Heart Disease

- ♥ Education and Training:
  - ♥ HRSA shall fund the development of, in collaboration with public health and health care professional organizations and families, appropriate education and training materials for families and public health and health care professionals relevant to the screening and treatment of CCHD

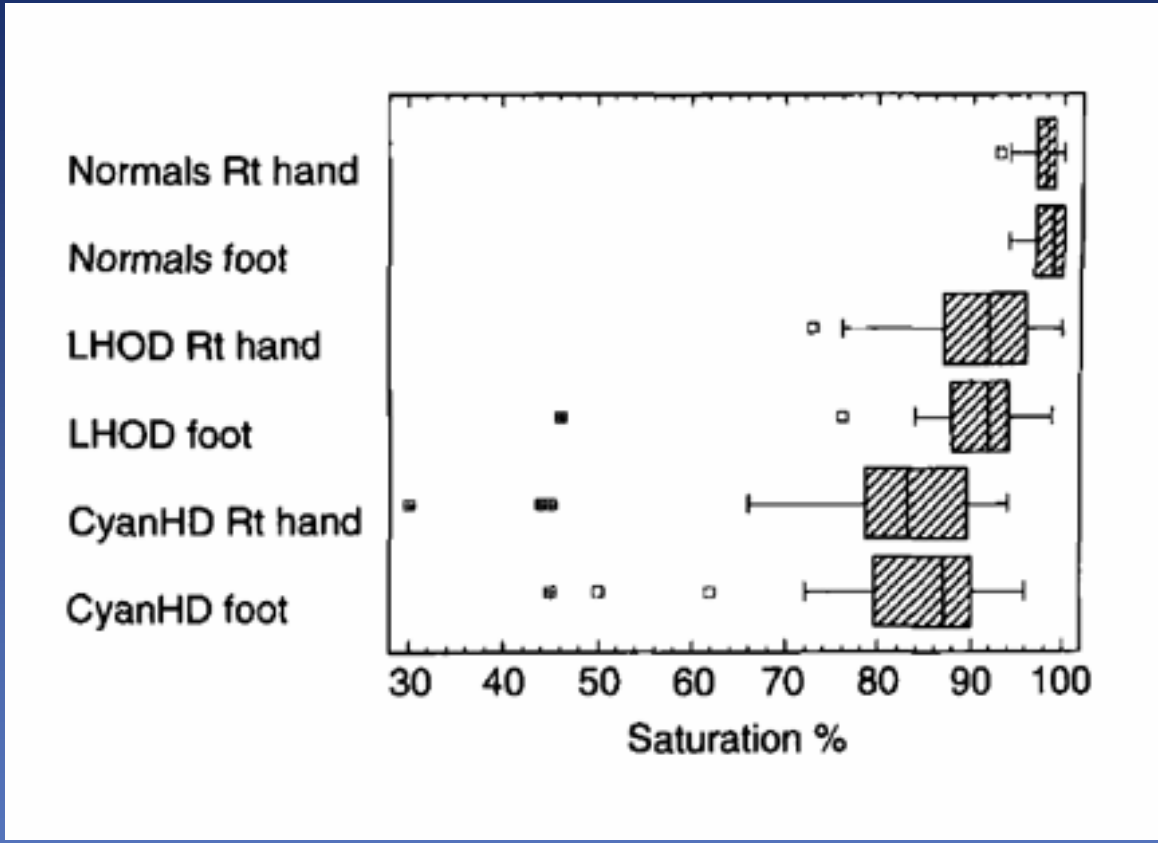
Why Screen

- Critical CHD has a higher frequency than other conditions that are universally screened for in the newborn nursery, including hypothyroidism and phenylketonuria
- Screening for critical CHD costs less than other universal newborn screenings
- Pulse Oximetry is an easy test to do, is painless, and a non-invasive way of measuring the oxygen saturation of hemoglobin in the arterial blood (the "5th" vital sign)

Pulse Oximetry



O_2 saturation values in patients with CCHD



de-Wahl Grannelli, A: Acta Paediatrica 2005;94:1590

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

Shakila Thangaratinam, Kiritrea Brown, Javier Zamora, Khalid S Khan, Andrew K Ewer

- Critical defects
  - All lesions that are duct dependent and required surgery in first 28 days after birth
- 552 studies, 13 primary studies were eligible for inclusion, 229,421 newborns were included
- Variability in studies included – inclusions of antenatal dx (4/12), timing of oximetry (5/12 less than 24 hours), foot alone (60%) or in conjunction with right hand, length of f/u.

	Limb	Antenatal diagnosis of CHD	Test timing	Total	True positive	False positive	False negative	True negative	Sensitivity (%; 95% CI)	Specificity (%; 95% CI)	Likelihood ratio positive (%; 95% CI)	Likelihood ratio negative (%; 95% CI)	False-positive rate (%; 95% CI)
Meberg et al (2008)[30]	Foot only	Excluded	<24 h	50 008	27	297	8	49 676	77·1% (59·9–89·6)	99·4% (99·3–99·5)	129·8% (104·9–160·6)	0·23% (0·13–0·43)	0·6% (0·5–0·7)
Bakr et al (2005)[25]	Foot and right hand	Excluded	>24 h†	5211	3	2	0	5206	100·0% (29·2–100·0)	100% (99·9–100·0)	1823·1% (500·1–6646·1)	0·13% (0·01–1·67)	0% (0–0·1)
Arlettaz et al (2006)[24]	Foot only	Included	<24 h	3262	12	12	0	3238	100·0% (73·5–100·0)	99·6 % (99·4–99·8)	250·1% (142·3–439·5)	0·04% (0·01–0·59)	0·4% (0·2–0·6)
Sendelbach et al (2008)[26]	Foot only	Excluded	<24 h	15 233	1	24	0	15 208	100·0% (2·5–100·0)	99·8 % (99·8–99·9)	466·3% (191·0–1138·5)	0·25% (0·02–2·8)	0·2% (0·1–0·2)
Reich et al (2003)[31]*	Foot and right hand	Excluded	>24 h†	2114	0	4	0	2110	..	99·8% (99·5–99·9)	..	..	0·2% (0·1–0·5)
Koppel et al (2003)[29]	Foot only	Excluded	>24 h	11 281	3	1	2	11 275	60·0% (14·7–94·7)	100·0% (100·0–100·0)	6765·6% (839·8–54 506·3)	0·40% (0·14–1·17)	0% (0·0–0·0)
Rosati et al (2005)[34]	Foot only	Excluded	>24 h	5292	2	1	1	5288	66·7 % (9·4–99·2)	100·0% (99·9–100·0)	3526·0% (424·6–29 282·9)	0·33% (0·07–1·70)	0% (0·0–0·1)
Richmond et al (2002)[32]	Foot only	Included	<24 h	5626	8	56	1	5561	88·9% (51·8–99·7)	99·0% (98·7–99·2)	89·2% (62·9–126·3)	0·11% (0·02–0·71)	1% (0·8–1·3)
de Wahl Granelli (2009)[16]	Foot and right hand	Excluded	>24 h†	39 821	19	68	10	39 724	65·5% (45·7–82·1)	99·8% (99·8–99·9)	383·4% (268·8–546·9)	0·35% (0·21–0·57)	0·2% (0·1–0·2)
Riede (2010)[33]	Foot only	Excluded	≥24 h	41 442	14	40	4	41 384	77·8% (52·4–93·6)	99·9% (99·9–99·9)	805·5% (542·0–1197·0)	0·22% (0·09–0·53)	0·1% (0·1–0·1)
Ewer et al (2011)[17]	Foot and right hand	Included	<24 h	20 055	18	177	6	19 854	75·0% (53·3–90·2)	99·1% (99·0–99·2)	84·9% (64·6–111·6)	0·25% (0·13–0·50)	0·9% (0·8–1·0)
Kawalec et al (2006)[28]	Foot only	Excluded	≥24 h	27 200	7	13	1	27 179	87·5% (47·3–99·7)	100·0% (99·9–100·0)	1830·2% (1001·2–3345·9)	0·13% (0·02–0·78)	0% (0·0–0·1)
Hoke et al (2002)[27]*	Foot and right hand	Included	<24 h	2876	4	53	0	2819	100·0% (39·8–100·0)	98·2% (97·6–98·6)	48·3% (32·6–71·7)	0·10% (0·01–1·40)	1·8% (1·4–2·4)
Summary estimate	..	..	..	229 421	..	..	..	..	76·5% (67·7–83·5)	99·9% (99·7–99 9)	549·2% (232·8–1195·6)	0·24% (0·17–0·33)	0·14% (0·06–0·33)

CHD=congenital heart defect. *Studies by Hoke and colleagues and Reich and colleagues excluded from the analysis. †Mean age at testing >24 h after birth.

Table: **Accuracy estimates of primary studies for pulse oximetry in the detection of critical congenital heart defects in newborn babies**

Timing and method of testing affects false positive rate

	Sensitivity	Specificity	False pos rate
Test < 24 hours Foot only	88.9-100%	89.2-99.6	0.4- 1%
Test < 24 hours Hand and foot	60-100%	99.4-100%	0-0.6%
Test > 24 hours Foot only	77-100%	98.2-99.1	0.9-1.8%
Test > 24 hours Hand and foot	65.5-100%	99.8-100%	0-0.2%

Variability in whether prenatal dx was included

Pulse oximetry as screening method

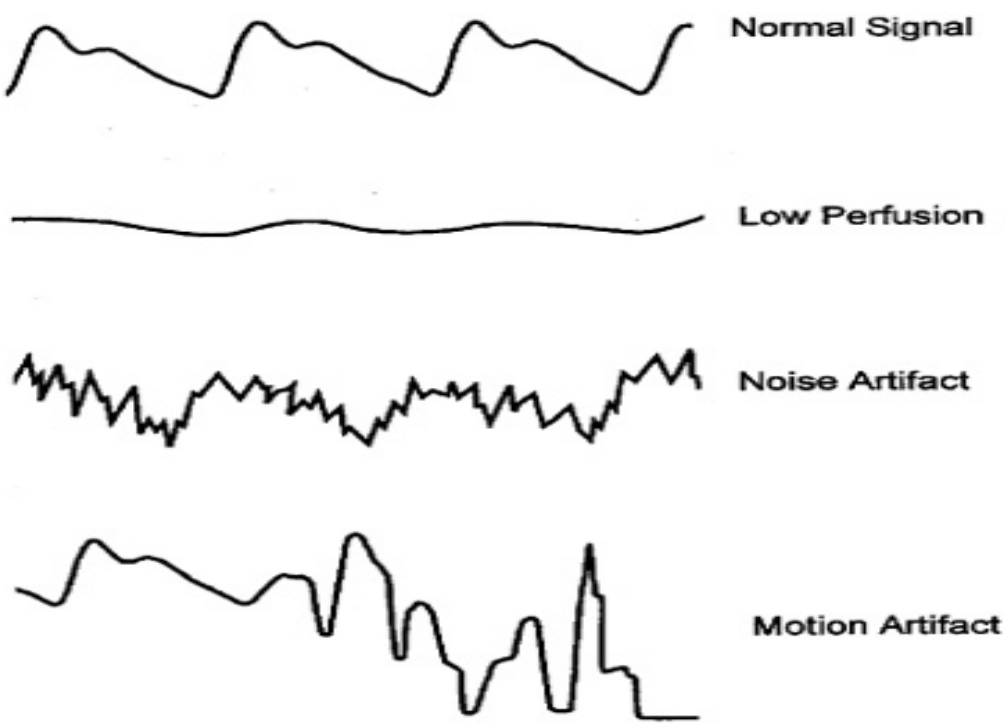
- Pulse oximetry measures the amount of O_2Hgb in the arterial blood
- Based on differential absorption of O_2Hgb and RHgb
- Coupled with ability to separate pulsatile from non-pulsatile components
- Non-invasive and painless
- Accurate with newer generation oximeters
- “Motion resistant” (SET) technology
- Fast (<2 min) and reliable
- Inexpensive (~ $4 per baby)
- Peripheral perfusion index (PPI)

Oxygen Sat is as simple as this

Pulse Oximetry

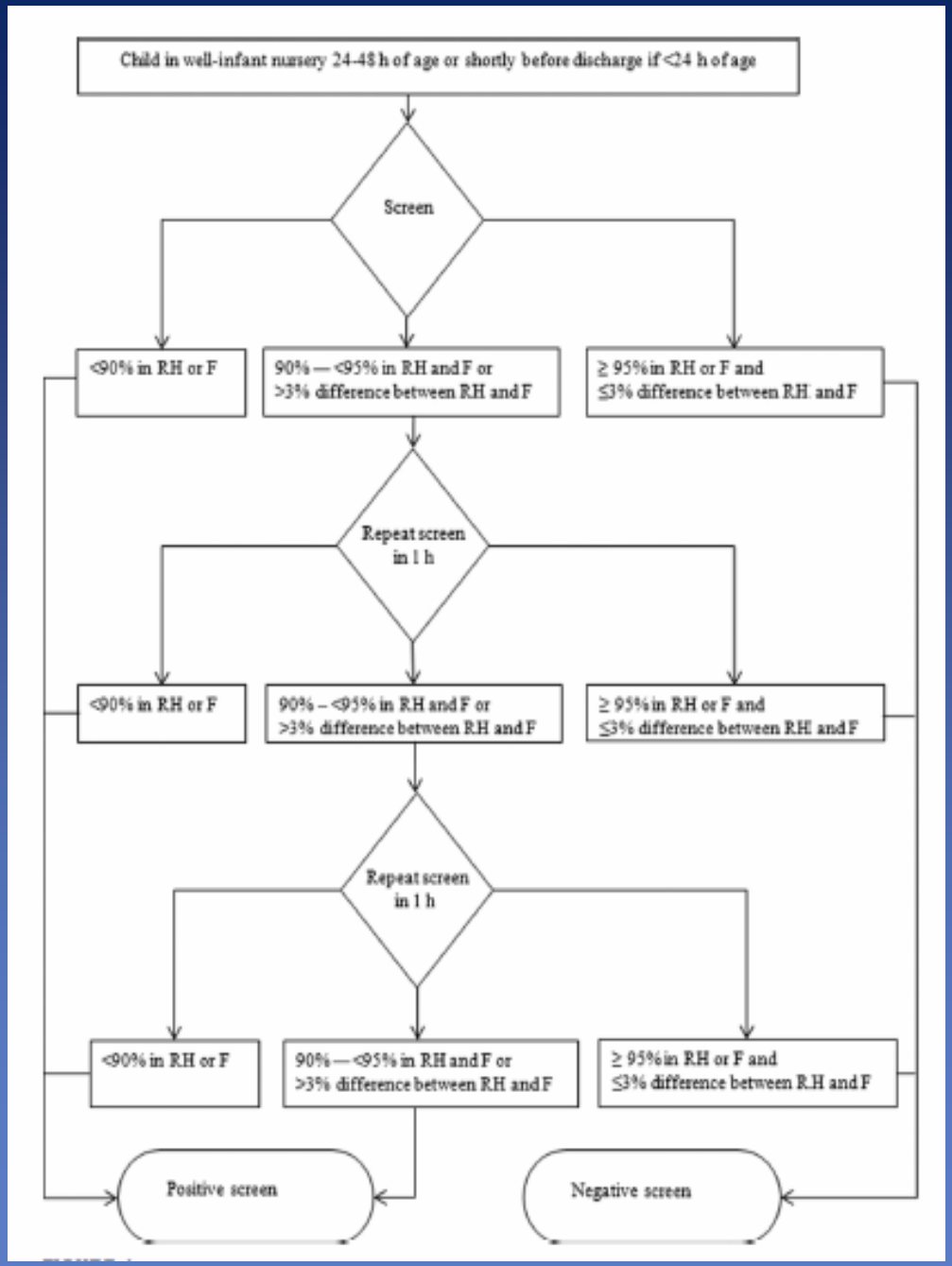
- Perform after 24hrs of age
- Infant should be awake
- Place Massimo probe on right hand followed by either foot. (performed either in parallel or in sequence)
- Obtain saturation and follow algorithm
- Remember to make sure you have a strong consistent pleth for accurate results

Pleth: the good, the bad, and the ugly



Pulse Oximetry

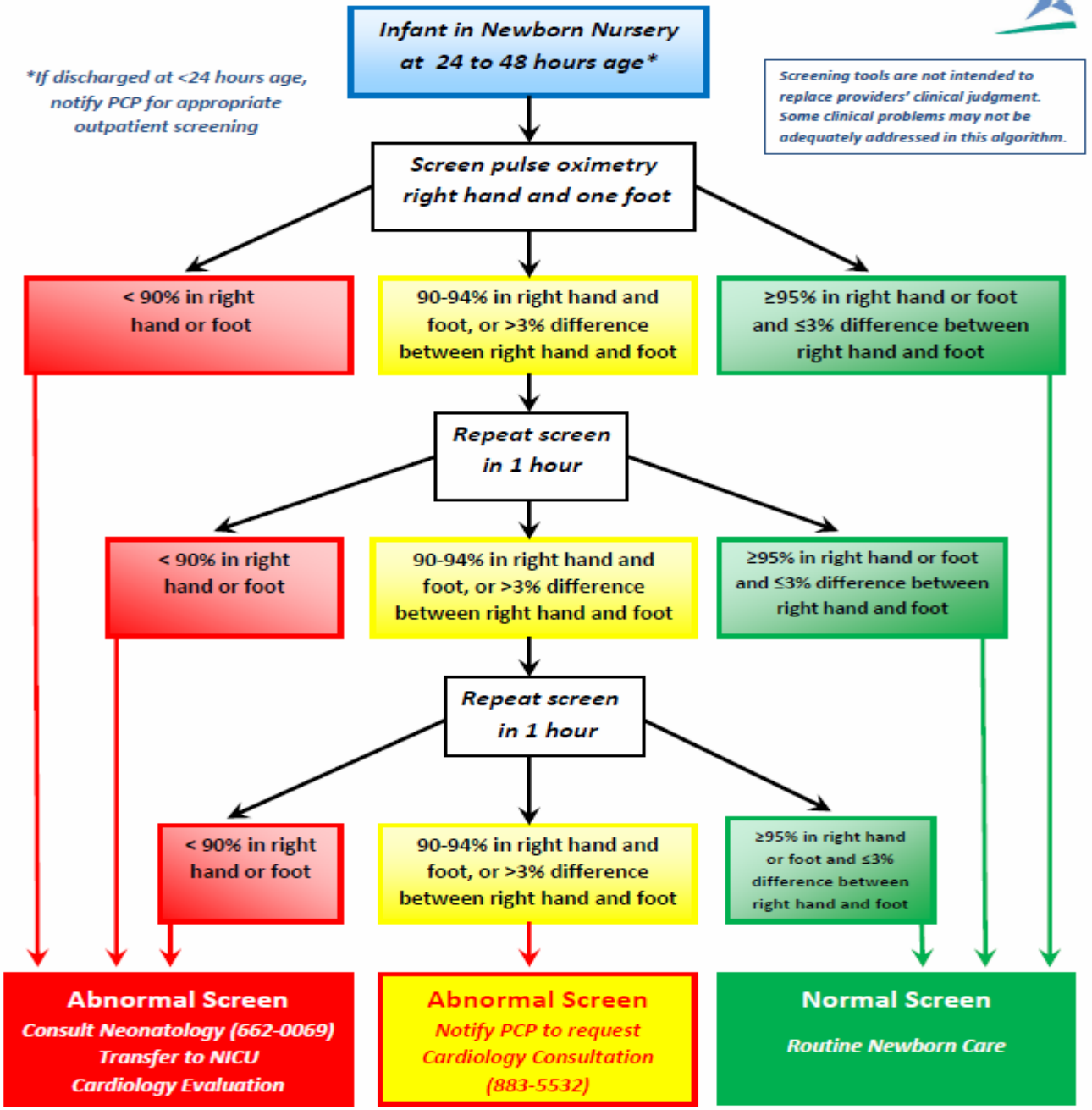
- Research has been conducted nationally and internationally to determine standards for best-practice for screening of critical CHD using pulse oximetry.
- Research shows that the highest sensitivity (true positives) and highest specificity (true negatives) is associated screening the right hand and one foot, using a cut-off of 95% or a 3% difference between the two
- Best outcomes are found when physical examination is paired with pulse oximetry screening.


Child in well-infant nursery 24-48 h of age or shortly before discharge if <24 h of age
Screen
<90% in RH or F
90% — <95% in RH and F or >3% difference between RH and F
≥ 95% in RH or F and ≤3% difference between RH and F
Repeat screen in 1 h
<90% in RH or F
90% – <95% in RH and F or >3% difference between RH and F
≥ 95% in RH or F and ≤3% difference between RH and F
Repeat screen in 1 h
<90% in RH or F
90% — <95% in RH and F or >3% difference between RH and F
≥ 95% in RH or F and ≤3% difference between RH and F
Positive screen
Negative screen

MMC Algorithm

Newborn Pulse Oximetry Screening

The Barbara Bush Children's Hospital
At Maine Medical Center



Educational Materials

Critical Congenital Heart Disease Screening Program

The Barbara Bush Children's Hospital
At Maine Medical Center

Educational Materials

Heart Smart Videos

- Available in Spanish, Simplified Chinese, Russian, French and Arabic
- Grant Funded from **Baby's First Test**

Video for Providers:
http://www.youtube.com/watch?v=Lif7kSgHfkw

Video for Families:
http://www.youtube.com/watch?v=o2CHeMRNdGg

Abnormal?

Nl Abn

- Right hand 88%; foot 89%?
- Right hand 95%; foot 91%?
- Right hand 97%; foot 93%?
- Right hand 98%; foot 95%?
- Right hand 96%; foot 94%?

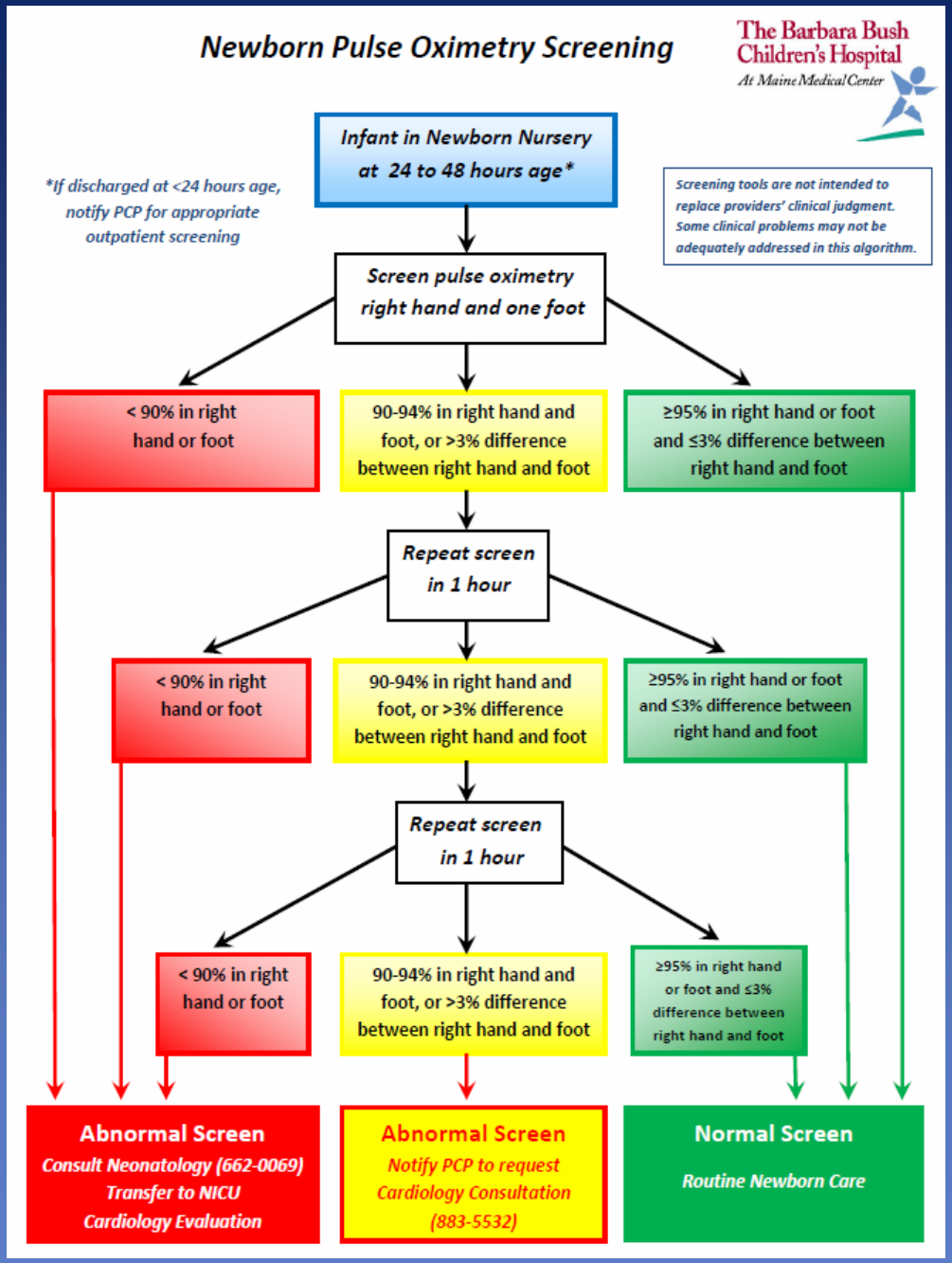
Abnormal?

	Nl	Abn
• Right hand 88%; foot 89%?		X
• Right hand 95%; foot 91%?		X
• Right hand 97%; foot 93%?		X
• Right hand 98%; foot 95%?	X	
• Right hand 96%; foot 94%?		X

Nl

Abn

- Right hand 88%; foot 89%?
- Right hand 95%; foot 91%?
- Right hand 97%; foot 93%?
- Right hand 98%; foot 95%?
- Right hand 96%; foot 94%?



Abnormal Result

- Oxygen Sat <90% no need to repeat
  - infant transferred to a NICU for evaluation
- O2 sat 90%-94% in both extremities: 3 measures 1 hour apart (algorithm) OR
- >3% difference in O2 sat between Rt hand and foot on 3 measures 1hr apart

Keep in mind- infant may NOT have heart disease!

Abnormal?

	Nl	Abn
• Right hand 88%; foot 89%?		X
• Right hand 95%; foot 91%?		X
• Right hand 97%; foot 93%?		X
• Right hand 98%; foot 95%?	X	
• Right hand 96%; foot 94%?		X

Abnormal Result

- Oxygen Sat <90% no need to repeat
  - infant transferred to a NICU for evaluation
- O2 sat 90%-94% in both extremities: 3 measures 1 hour apart (algorithm)
- OR >3% difference in O2 sat between Rt hand and foot on 3 measures 1hr apart

Keep in mind- infant may NOT have heart disease!

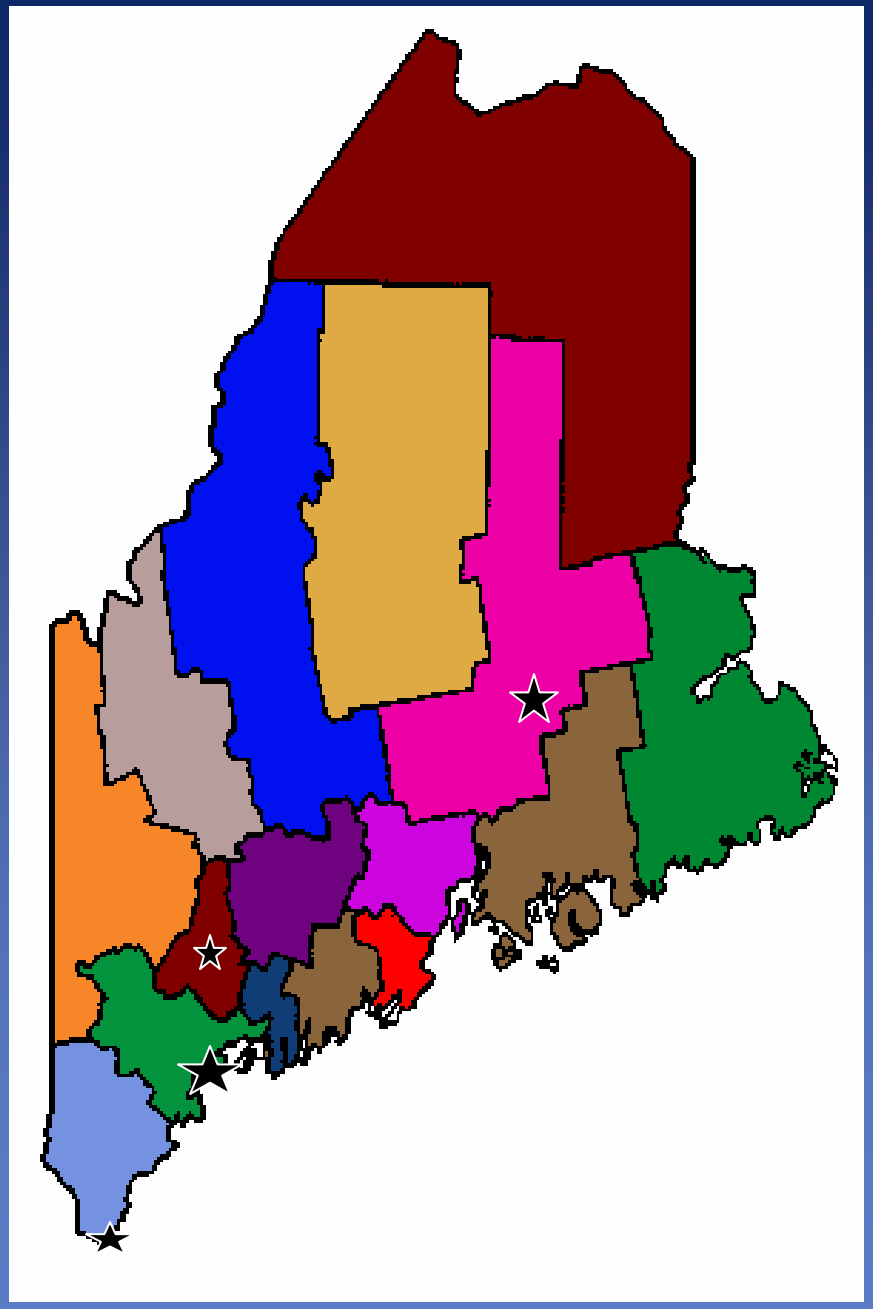
Abnormal result

- Notify Primary Care Provider
- Physical exam
  - Auscultation of heart and lung sounds
  - 4 extremity blood pressures
  - Capillary refill
  - Femoral pulses
  - ? Chest X-Ray?
  - ? Continuous saturation monitoring?
  - ? Blood Gas?

Abnormal result, “What’s next?”

- Does the baby need to stay in NBN as opposed to Mom’s room?
- Will baby need an ECHO?
- Will the baby be transported or discharged?
- Who tells the parents?

Maine sites with Pediatric Cardiologists



HRSA Grant

- 3 years
  - June 1, 2012-May 31, 2015
- Total National funding
  - 2,100,000.00
  - 7 regional screening programs
    - New England Genetics Collaborative
    - ME, NH, VT, RI, CT
  - 300,000 per region
- Personnel-data entry, analysis, education
- Meetings-twice per year

HRSA Grant

- Goals and Objectives:
  - Develop a plan to incorporate CCHD screening methods at birthing facilities
  - Develop guidelines to collect and report results
  - Establish guidelines for screening follow-up and reporting
  - Develop education programs for providers and families

HRSA Grant

- Goals and Objectives (cont.):
  - Develop methods for quality assurance, outcomes analyses (including costs), and public health monitoring
  - Establish baseline data for each screening facility and develop ongoing data collection, analysis, and reporting methods
  - Establish state level health information exchange systems (requires legislation in Maine)
  - Prepare a project evaluation that includes next steps and identified best practices.

Legislation
LD 460

Maine Statistics Estimations

- Approximately 13,000 babies per year
- Incidence of 8:1000
- Anticipate 104 cases per year of CHD
- Of those- 25 cases per year of CCHD
  - Prenatal Diagnosis (~10-16)
  - Positive Screens (~ 13-20)
  - Undetected by Screening (~ 1-4)
- False Positives- (~25 babies per year)

Maine Implementation and Data

- Tertiary Care
  - EMMC (January 2012 implemented)
    - No CCHD diagnosed by pulse oximetry
      - 1 false negative (coarctation)
      - others ill or prenatally diagnosed
  - MMC (May 2012 implemented)
    - No CCHD diagnosed by pulse oximetry
      - ~1800 babies screened
      - Three abnormal screens
        - » ASD, PFO, false positive
      - 2 parent refusals

Status of Implementation

- Already Implemented:
  - Farmington
  - Waldo County General
  - Bridgton
  - Mercy
  - Calais
  - Cary
  - CMMC
  - Down East Community
  - Goodall
  - Houlton
  - Maine General-both campuses
  - Penobscot Bay
  - SMMC
  - St. Mary's
  - The Aroostook Medical Center
  - York
- In Process:
  - Mayo Regional
  - Mid Coast
  - Miles
  - Mount Desert
  - Northern MMC
  - Penobscot Valley
  - Redington Fairview
  - Rumford
  - Stephens Memorial

Approximately 2/3 of babies born in Maine are currently screened by pulse oximetry

Screening Stories from the Field

- Centers screening
  - both arms and neither leg
  - Q 15 minutes after birth X 1 hour and then one arm at 24 hours
  - results with greater than 3% difference evaluated by pediatric provider but not referred to cardiology
  - after Prenatal diagnosis of CHD already made

Points to leave with

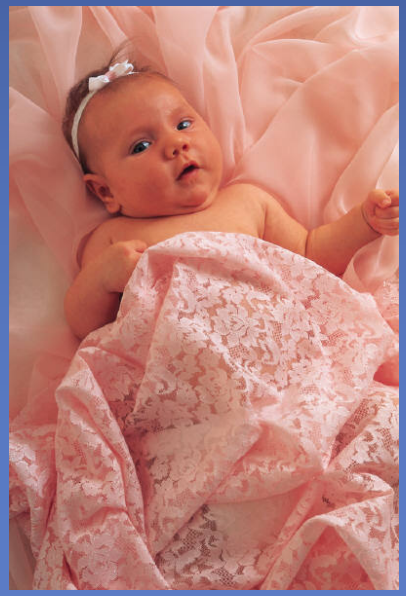
- Newborn saturation screening is happening in ME; most centers following the AAP guideline
- Clinical examination alone misses critical CHD
- Oximetry is stable and reliable
- False positive rates are low (lower than false positive rate based on physical exam)
- Data to support this is strong
- Some lesions still will not be detected

If you remember nothing else…

- Right hand and either foot, over 24 hours
- Know the cutoffs (< 90, or ≥ 95%)
- Pay attention to the 3% difference
- Remember your exam is still important
  - Auscultation; femoral pulses (4 ext if needed)
  - CXR
  - Keep in mind, "*it might not be the heart*"
  - EKG- likely not necessary unless requested by pediatric cardiology

Selected References

- www.childrensnational.org/pulseox
- Strategies for Implementing Screening for Critical Congenital Heart Disease, Kemper, MD et al. *Pediatrics* 2011 Nov;128(5): e1-9
- Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies. Lancet 2012. Thangaratinam et al.
- Endorsement of Health and Human Services Recommendation for Pulse Oximetry Screening for Critical Congenital Heart Disease. AAP Section on cardiology and cardiac surgery executive committee, January 2012.



Questions?