MAWS Fall Conference 2013

Pulse Oximetry Screening for Critical Congenital Heart Disease

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Disclosures: none

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The new recommendation: Fall 2011

- All newborns should be screened for critical congenital heart disease by pulse oximetry prior to discharge from the hospital.
- Primary care providers will need to develop mechanisms for screening newborns who missed being screened at birth.
- Recommended by:
  - Health and Human Services
  - American Academy of Pediatrics
  - American College of Cardiology
  - American Heart Association

Kemper AR et al, *Pediatrics* 2011

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The new recommendation: Fall 2011

- "The work group chose not to focus on out-of-hospital births, which raise challenging coordination-of-care issues, which will be addressed in the future."

Kemper AR et al, *Pediatrics* 2011
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Case #1

- 4 day old term male
  - Uncomplicated pregnancy
  - Home at 2 days of age
- Presented to community hospital ER in shock at 4 days
  - Profound metabolic acidosis (pH 6.99)
  - Transferred to children's hospital ICU
- Echo revealed hypoplastic left heart syndrome
  - PGE started
- Evidence of liver/kidney injury but recovered→successful staged palliation
- Can't we do better?

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Can we do better?

- ~40% of critical CHD is detected prenatally in WA
- The physical exam has limited sensitivity for critical CHD
  - Often no pathologic murmur
  - It is hard to see cyanosis!

SaO_2	% time lips rated cyanotic
>90%	28%
85-89%	55%
80-84%	60%
75-79%	74%
<75%	94%

Goldman J Peds 1973

- To do better, we need new methods of screening
- Pulse oximetry is now recommended as the method

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Overview

- "Critical" CHD
  - What is it?
  - Review physiology
  - Association with hypoxemia
- Pulse oximetry as a screening tool
- Implementation
  - Obtaining reliable readings
  - Screening algorithm interpretation
  - Management of abnormal screening results
  - Examples

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What is "Critical" CHD?

- CHD requiring invasive intervention in 1st month of life
  - Overall incidence ~1/1000 live births
- Lesions targeted by the expert panel:
  - Hypoplastic left heart syndrome
  - Pulmonary atresia
  - Tetralogy of Fallot
  - Tricuspid atresia
  - Transposition of the great arteries
  - Truncus arteriosus
  - Total anomalous pulmonary venous connection
- Milder forms of CHD are *not* targeted; unlikely to be identified

Aamir T *Acta Pediatrica* 2007, Wren C *Arch Dis Child Fetal Neonatal* 2008, DeWahl Granelli *BMJ* 2009, Riehle-Colarusso T *Congenit Heart Dis* 2007.

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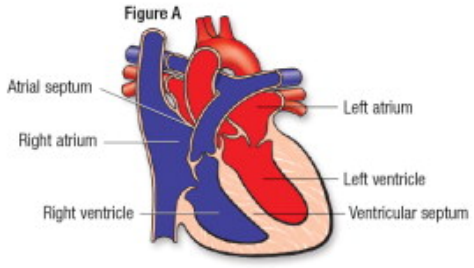
Review of normal cardiac anatomy



www.heart.org

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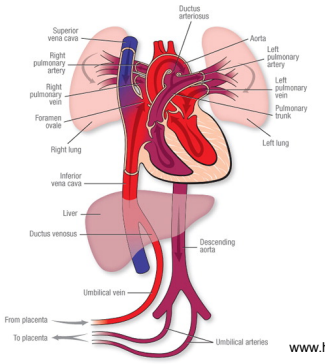
Review of the fetal circulation



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The physiology of transition: major changes after birth

Change	Time course	Lesions presenting
Increased pulmonary blood flow→increased pulmonary venous return	Seconds-minutes	Obstructed pulmonary venous return
Ductal closure	Hours-days	Ductal dependent lesions
Continuing drop in pulmonary vascular resistance (PVR)	Weeks-months	Shunt lesions

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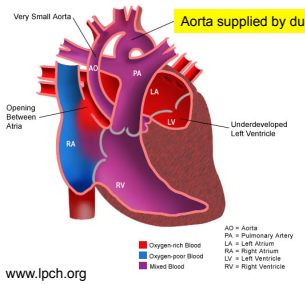
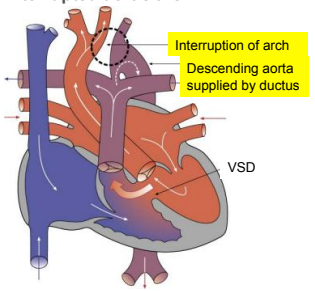
Ductal dependent systemic blood flow

Hypoplastic left heart syndrome



www.lpch.org

Interrupted aortic arch



www.pediatriccardiacinquest.mb.ca

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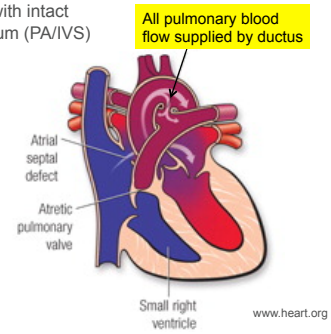
Ductal dependent pulmonary blood flow

Pulmonary atresia with intact ventricular septum (PA/IVS)



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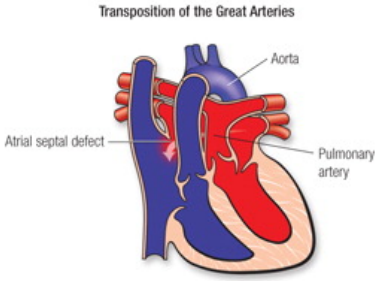
Transposition of the Great Arteries

- Two parallel circulations
- Survival is dependent on mixing (bidirectional shunting)
- Mixing is best at the atrial level, but ductal patency can assist with mixing.



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Critical CHD and Hypoxemia

- Do all forms of critical CHD have similar oxygen saturations?

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Critical CHD and Hypoxemia

- Do all forms of critical CHD have similar oxygen saturations? *NO*
- In any lesion where there is *complete mixing* of red and blue blood, the oxygen saturation depends on the ratio of pulmonary to systemic blood flow

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Critical CHD and Hypoxemia

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Critical CHD and Hypoxemia

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Critical CHD and Hypoxemia

Lesion	Typical SpO_2 with ductus open
Ductal-dependent systemic blood flow	90's
Ductal-dependent pulmonary blood flow	80's
D-Transposition of the great arteries with good mixing with poor mixing	 80's Low

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Differential Cyanosis

- Oxygen saturations differ between extremities
- Part of the aorta supplied by the LV
- Another part supplied by the RV

} One of these is via the ductus

- RUE > foot: critical arch obstruction
- RUE < foot: transposition

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Screening

- How common is "missed critical CHD"?
- How well does pulse oximetry perform as a screening test?
- What is the best time window for screening?
- What are normal values for pulse oximetry in newborns?

Chapter on Screening from *Epidemiology in Medicine*, Hennekens CH and Buring JE, 1987

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How common is "missed" critical CHD?

- Critical CHD
  - ~1/1000 live births
  - Prenatal diagnosis ~40% (WA)
- Diagnosis after initial discharge from hospital
  - ~1/4000 to 1/14,000 live births
- Diagnosis at autopsy
  - Becoming more uncommon
  - ~1-2/100,000 live births

Aamir T *Acta Pediatrica* 2007, Wren C *Arch Dis Child Fetal Neonatal* 2008, DeWahl Granelli *BMJ* 2009, Riehle-Colarusso T *Congenit Heart Dis* 2007, Chang RK *Circulation* 2007 (abstract)

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"Missed" critical CHD compared to other newborn screening targets

Disorder	Incidence (per # live births)
Congenital hearing loss	1-2/1000
Congenital hypothyroidism	1/3000
Sickle cell disease	1/3700
"Missed" critical CHD	1/4000-1/14,000
Phenylketonuria	1/14,000
Congenital adrenal hyperplasia	1/19,000
Galactosemia	1/53,000
Biotinidase deficiency	1/62,000
Maple syrup urine disease	1/230,000
Homocystinuria	1/343,000

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Pulse oximetry as a screening tool: how well does it work?

- >13 studies examining this question
  - Different cutoffs, timing, etc.
- DeWahl Granelli et al, *BMJ* 2009
  - Used algorithm recommended in guideline
  - Screened 38,429 Swedish newborns

	CHD no	CHD yes	calculations
Pass screen	38259	11	
Fail screen	65	22	PPV 25% 87 = 1/440
Incomplete screen	72		
Calculations		Sensitivity 67%	

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Pulse oximetry as a screening tool: how well does it work?

- All false negatives had ductal dependent systemic blood flow
  - Oximetry detected 10/20 (50%)
  - 4/10 missed by oximetry had weak/impalpable femoral pulses at day 1-4 of life
- No case of ductal dependent pulmonary blood flow or transposition was missed (n=9)
- Pulse oximetry performed much better than PE
- Pulse oximetry *plus* PE had the highest sensitivity for critical CHD (83%)

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Pulse oximetry as a screening tool: how well does it work?

- DeWahl Granelli et al, *BMJ* 2009, cont'd:
  - ~50% of false positives had non-ductal dependent CHD, sepsis or pulmonary disease
- Outcomes:
  - Screening decreased:
    - discharge to home without diagnosis (8% vs 28%)
    - severe acidosis at diagnosis (12% vs 33%)
  - Babies identified before d/c had lower surgical mortality

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Timing of Screening

- Recommended:
  - Between 24 & 48 hours of age
- Needs to be optimized
  - Too late—miss too many babies who get sick
  - Too early—too many false positives

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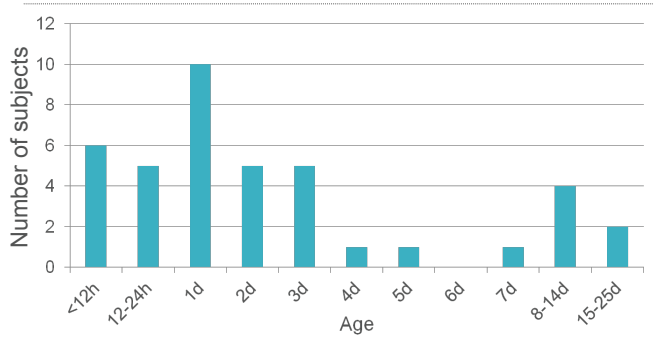
What is the time window for detection of critical CHD by screening?



Schultz AH et al *Pediatrics* 2008

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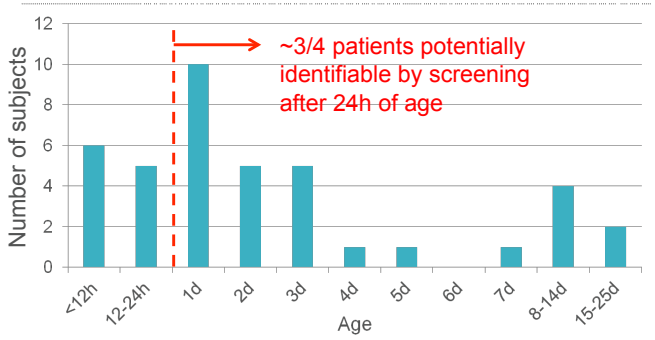

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What is normal for pulse oximetry in the newborn?

- Normal SpO_2 values in healthy term newborns

Age	Median SpO_2
1 minute	70's
5 minutes	80's
10 minutes	Low 90's
Admission to newborn nursery	97%
24 hours	97%

- However, 4-8% of healthy term babies have significantly lower SpO_2 in the first 24 hours of life (median 92%)
  - Most common on admission to newborn nursery

Levesque BM *Pediatric Pulmonol* 2000, O'Brien LM *Arch Dis Child Fetal Neonatal Ed* 2000, Dimich I *Can J Anaesth* 1991

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Timing of Screening

- Thangaratinam et al Lancet 2012
  - Meta-analysis of 13 studies of pulse oximetry screening
  - 229,421 patients
  - Screening at:
    - <24h false positive rate: 1/200
    - >24h false positive rate: 1/2000
  - However, "<24h" was typically between 2 and 12 hours of age
  - Little data to guide what false positive rate will be at 18-24h

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What results can you expect from pulse ox screening?

- ~1/500 - 1/1000 babies will fail the screen
  - ~20-25% have critical CHD
  - ~45-50% have other conditions worth identifying
- Pulse oximetry screening improves detection of critical CHD and can reduce surgical mortality
- False positives increase if screening is done before 24h
- Optimal timing 24-48 hours of age
- Screening typically takes 4-7 minutes

Kemper et al. Pediatrics 2011, de-Wahl Granelli et al BMJ 2009, Thangaratinam S et al Lancet 2012

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What results can you expect from pulse ox screening?

- Screening has an overall sensitivity of 60-75% and thus a normal screen does *not* rule out critical CHD
- Pulse oximetry reliably detects:
  - Transposition of the great arteries
  - Lesions with ductal dependent *pulmonary* blood flow
- Pulse oximetry less reliably detects:
  - Lesions with ductal dependent *systemic* blood flow
  - Sensitivity ~50%
  - Pay attention to femoral pulses on exam

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Obtaining reliable readings

- Use a high quality pulse oximeter
  - FDA approved for neonatal use
  - Reports functional (not fractional) oxygen saturation
- Use the manufacturer recommended probe
- Use taped, not clamped sensors
- The baby should be awake, warm and quiet, not feeding
- Block ambient light from probe
- The pulse oximeter should be picking up all heartbeats
  - Look at the bouncing bar/waveform
  - Check against HR/pulse
  - Motion is the usual culprit if not picking up all beats

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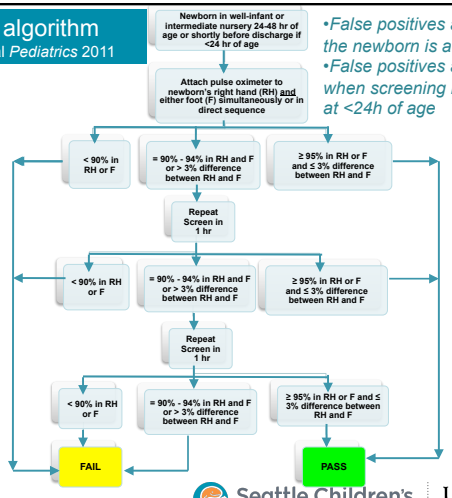
Screening algorithm

Kemper AR et al *Pediatrics* 2011



•False positives are reduced if the newborn is awake
•False positives are increased when screening is performed at <24h of age

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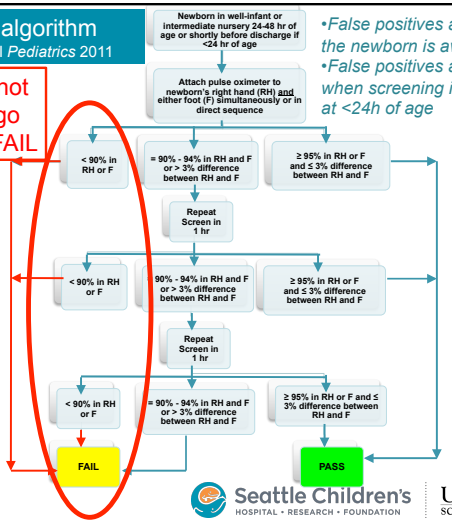
Screening algorithm

Kemper AR et al *Pediatrics* 2011

<90%: do not pass Go…go directly to FAIL



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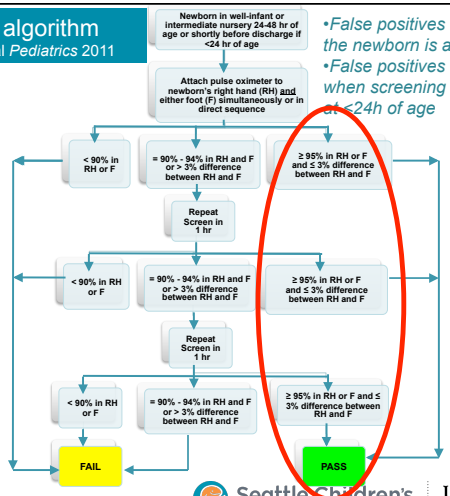

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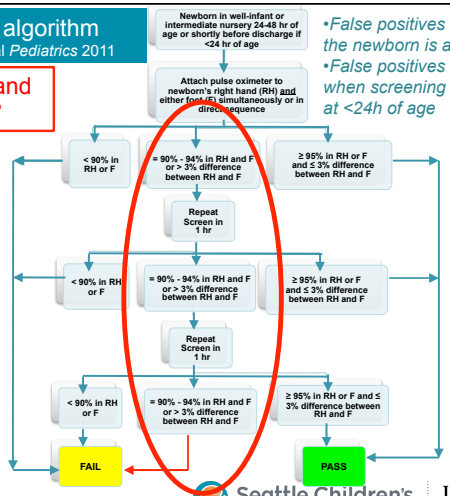
Screening algorithm

Kemper AR et al *Pediatrics* 2011

"3 strikes and you're out"



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- *False positives are increased when screening is performed at <24h of age*

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Pulse Ox Screening is **FAILED** if either of the following are true:

1. Any correctly obtained oxygen saturation is < 90% OR
2. The patient does not pass on 3 consecutive trials separated by 1 hour, either:
   a) Oxygen saturation is <95% in both extremities
   b) There is a > 3% difference in oxygen saturation between the right hand and either foot.

Cutoffs for high altitude not yet determined

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So the baby failed the screen… now what?

- Notify responsible care provider
  - Perform complete clinical evaluation
- If no explanation for hypoxemia, echo is indicated.
  - Discuss with Pediatric Cardiologist prior to echo (strongly recommended) .
  - No echo on-site→consider transfer.
  - A failed screen should be resolved *prior to discharge*.

Kemper AR et al, *Pediatrics* 2011

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How can Seattle Children's help?

- SCH Cardiologists are available 24/7
  - Discuss the clinical situation of newborns with positive screens
  - Facilitate echocardiography or transfer
  - Physician operator 206-987-7777
  - Or paging operator 206-987-2000
- SCH Cardiologists read echocardiograms for multiple hospitals in Washington on a regular basis
  - Direct digital image transfer to SCH

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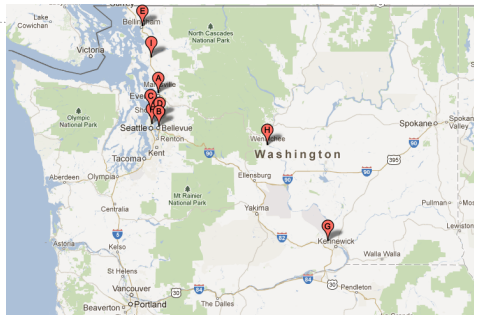
Where can you get an echo? SCH telemedicine sites

	Telemedicine/outside echo sites
A	Providence Everett Medical Center
B	Overlake Hospital
C	Swedish Hospital Edmonds (CD only)
D	Evergreen Hospital
E	St Joseph Bellingham
F	UW Medical Center
G	Kadlec Regional Medical Center
H	Central Washington Wenatchee
I	Skagit Valley Hospital



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Resources

- Resources
  - http://www.seattlechildrens.org/healthcare-professionals/gateway/pulse-oximetry-screening-newborns/
  - Pulse ox tool (Children's Healthcare of Atlanta) http://pulseoxtool.com/index.php

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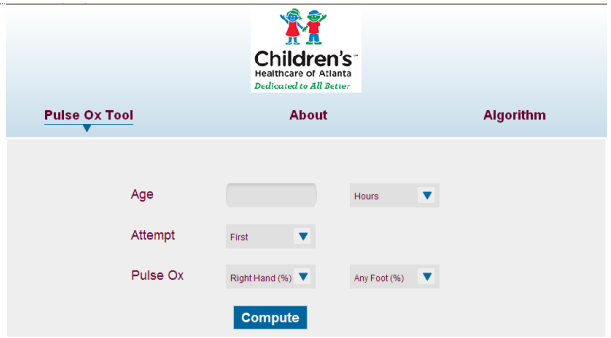
Resources



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Example 1

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	98%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 1

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	98%		
Either foot	96%		
Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 1

	Test #1	Test #2	Test #3
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Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
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Probe has been applied. Ready for Example 2

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Example 2

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	92%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 2

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	92%		
Either foot	90%		
Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 2

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	92%		
Either foot	90%		
Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 2

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1340	
Right hand	92%	92%	
Either foot	90%	89%	
Difference	2%	3%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 2

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1340	
Right hand	92%	92%	
Either foot	90%	89%	
Difference	2%	3%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Probe has been applied. Ready for Example 3

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot	92%		
Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot	92%		
Difference	2%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	94%	95%	
Either foot	92%	91%	
Difference	2%	4%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	94%	95%	
Either foot	92%	91%	
Difference	2%	4%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	94%	95%	95%
Either foot	92%	91%	92%
Difference	2%	4%	3%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 3

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	94%	95%	95%
Either foot	92%	91%	92%
Difference	2%	4%	3%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Probe has been applied. Ready for Example 4

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Example 4

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	88%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 4

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	88%		
Either foot	84%		
Difference	4%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 4

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	88%		
Either foot	84%		
Difference	4%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Probe has been applied. Ready for Example 5

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	98%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	98%		
Either foot	94%		
Difference	4%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	98%		
Either foot	94%		
Difference	4%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	98%	100%	
Either foot	94%	92%	
Difference	4%	8%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	98%	100%	
Either foot	94%	92%	
Difference	4%	8%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	98%	100%	99%
Either foot	94%	92%	93%
Difference	4%	8%	6%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 5

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	98%	100%	99%
Either foot	94%	92%	93%
Difference	4%	8%	6%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Probe has been applied. Ready for Example 6

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot			
Difference			
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot	93%		
Difference	1%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235		
Right hand	94%		
Either foot	93%		
Difference	1%		
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	94%	93%	
Either foot	93%	92%	
Difference	1%	1%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	
Right hand	94%	93%	
Either foot	93%	92%	
Difference	1%	1%	
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	94%	93%	93%
Either foot	93%	92%	91%
Difference	1%	1%	2%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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Example 6

	Test #1	Test #2	Test #3
Date/Time	6/7/12 1235	6/7/12 1335	6/7/12 1435
Right hand	94%	93%	93%
Either foot	93%	92%	91%
Difference	1%	1%	2%
Result	Pass Repeat Fail	Pass Repeat Fail	Pass Fail
Signature			

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A real case

- First child screened by a midwife after purchase of pulse oximetry equipment in 2011
- Term boy born to a 32 yo G_7P_5 mother
  - Uncomplicated pregnancy
  - No family hx of CHD
  - Refused 20 week anatomy scan
  - GBS+, received 4 doses IV Penicillin G due to PROM
  - Apgars 9^1,10^5

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A real case

- Home visit at ~36 hours
  - Pulse oximetry readings 84-93%
  - Tried resetting machine, wrapping baby to limit movement
  - "I was convinced I didn't know what I was doing"
  - Other VS normal, baby well appearing, reported to be feeding well
  - On exam, no murmur, lungs clear
- F/U visit at 3 ½ days
  - Pulse oximetry 85-92%
  - Otherwise baby seemed to be doing great
  - The midwife "was ready to send the machine back"

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A real case

- DOL #7
  - Seen by pediatrician
  - Back to birth weight, pink, active and alert
- DOL #10
  - Call from Mom to midwife, seeking advice
  - Baby refusing to eat, vomiting
  - Some family members ill
  - Midwife advised taking baby to ER
  - Family deferred, instead planned to see pediatrician the next day

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A real case

- DOL #11
  - Seen by pediatrician
  - Did not hear obvious cardiac abnormality on exam, but pulse oximetry still abnormal
  - Sent to ED by ambulance, from there transported to SCH emergently
  - Oxygen saturations in the 30-50s on arrival
  - Dx: a form of transposition of the great arteries
  - Underwent emergent balloon atrial septostomy for stabilization
  - Subsequently has undergone multiple cardiac surgeries

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Messages

- The pulse oximeter, if used properly, is better than your eye at picking up desaturation
- Some forms of complex CHD have no obvious findings on exam, believe it or not!
- Know how to use/troubleshoot your pulse oximeter, then take it seriously if readings are abnormal

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Other References and Resources

- Seattle Children's contacts
  - Amy Schultz, MD (Cardiology)
    amy.schultz@seattlechildrens.org
- CDC website link to CCHD screening resources:
  http://www.cdc.gov/ncbddd/pediatricgenetics/CCHDscreening.html
  - Parent handout

    http://www.cdc.gov/ncbddd/pediatricgenetics/documents/CCHD-factsheet.pdf

Other References and Resources

- AAP Guideline:
  - Kemper AR, Mahle WT, Martin GR, Cooley WC, Kumar P, Morrow WR, Kelm K, Pearson GD, Glidewell J, Grosse SD, Lloyd-Puryear M, Howell RR. Strategies for Implementing Screening for Critical Congenital Heart Disease. *Pediatrics.* 2011; 128:e1-e8.

    http://pediatrics.aappublications.org/content/early/2011/10/06/peds.2011-1317.full.pdf
- Children's National Medical Center pulse oximetry screening resources:
  http://www.childrensnational.org/PulseOx/

References and Resources

Other references:

- de-Wahl Granelli A, Wennergren M, Sandberg K, et al, Impact of pulse-oximetry screening on the detection of duct-dependent congenital heart disease: a Swedish prospective screening study in 39,821 newborns. *BMJ* 2009;338:a3037.
- Mahle WT, Newburger JW, Matherne GP, et al, Role of pulse oximetry in examining newborns for congenital heart disease: a scientific statement from the AHA and AAP. *Pediatrics* 2009;124(2): 823-836.

References and Resources

Other references:

- Riede FT, Worner C, Dahnert I, Effectiveness of neonatal pulse oximetry screening for detection of critical congenital heart disease in daily clinical routine: results from a prospective multicenter study. Eur J Pediatr 2010; 169(8):975-981.
- Thangaratianam S, Brown K, Zamora J et al, Pulse oximetry screening for critical congenital heart defects in asymptomatic newborn babies: a systematic review and meta-analysis. *Lancet* 2012 epub May 2 ahead of print.