Critical Congenital Heart Disease (CCHD) Screening

Office for Genetics and People with Special Health Care Needs,
Maryland Department of Health & Mental Hygiene
Prevention and Health Promotion Administration

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Leadership Team

- National Leadership Academy for the Public's Health - A Project of the Center for Health Leadership and Practice (CHLP), Public Health Institute (PHI) - Oakland, CA and funded by the Centers for Disease Control (CDC):
  - Cynthia Mueller, RN, BSN
  - Dianna Abney, MD, MPH
  - Carissa Baker-Smith, MD, MPH
  - Debbie Badawi, MD

Background- CCHD

- About 25% of congenital heart disease is considered “critical”
- Diagnosis now relies on prenatal diagnosis or neonatal signs/symptoms
- Delayed diagnosis of CCHD causes increased morbidity and mortality
- Pulse ox screening of newborns can improve detection of CCHD when combined with a thorough exam

2011 Legislation

- Required Advisory Council on Hereditary and Congenital Disorders to convene expert panel to evaluate and submit legislative report 12/31/11
- Required Maryland to follow recommendation of Secretary Sebelius

2011 Legislation

- Legislative report available at http://fha.dhmh.maryland.gov/genetics/docs/CCHDLegisRpt.pdf
- In September 2011, CCHD Screening was adopted to RUSP

Advisory Panel for CCHD Screening

- Carissa Baker-Smith, MD MPH
- Miriam Blitzer, PhD
- Charlene Bennett, RN
- Carrie Blout, MS, CGC
- Elizabeth Bradshaw, MSN, RN, CPN
- Joel Brenner, MD
- David Bromberg, MD
- Debbie Burke, RN
- Maria Cardona, MD
- Joann Cordeiro
- Anne Eder
- Julie Hoover-Fong, MD, PhD
- Renee Fox, MD
- Maureen Gilmore, MD
- Tanya Green, MS, CCC-A
- Carole Greene, MD
- Linda Grogan, RNC, BSN, MBA
- Debra Harper-Hill, RN
- Sandra Heeley, RN
- Kimberly Iafolla, MD
- Julie Kaplan, MD
- Edward Lawson, MD
- Gerard Martin, MD
- Neil Porter, MD
- Geoffrey Rosenthal, MD, PhD
- Ann Sober, RN, BSN
- Philip Spevak, MD
- Johnna Watson, RN, BSN
- Anika Wilkerson
- S. Lee Woods, MD, PhD
- Cynthia Mueller, RN, BSN
- Debbie Badawi, MD
- Dianna Abney, MD, MPH

Implementation

- Screening
- Evaluation of Abnormal Results
- Recording Results
- Surveillance

The Goal for CCHD Screening

- Identify those newborns with structural heart defects usually associated with hypoxia in the newborn period that could have significant morbidity or mortality early in life with closing of the ductus arteriosus or other physiologic changes early in life.

Screening

- All babies should be screened prior to discharge
- NICU's need to develop protocols for screening (Lakshminrusimha S et. al, e-Journal of Neonatology Research Volume 2, Issue 2, Spring 2012)
- performed by a health professional whose scope of practice includes performing pulse oximetry, including CNA's

Screening

When:

- 24-48 hours of age
- while the infant is awake and quiet if possible
- avoid performing pulse oximetry on a crying or cold infant

Where:

- quiet area
- with parent present to soothe and comfort the infant (if possible)

Screening

- Machines should be approved by the FDA for use in neonates.

- Pre and Post-ductal saturations needed (right hand and one foot).

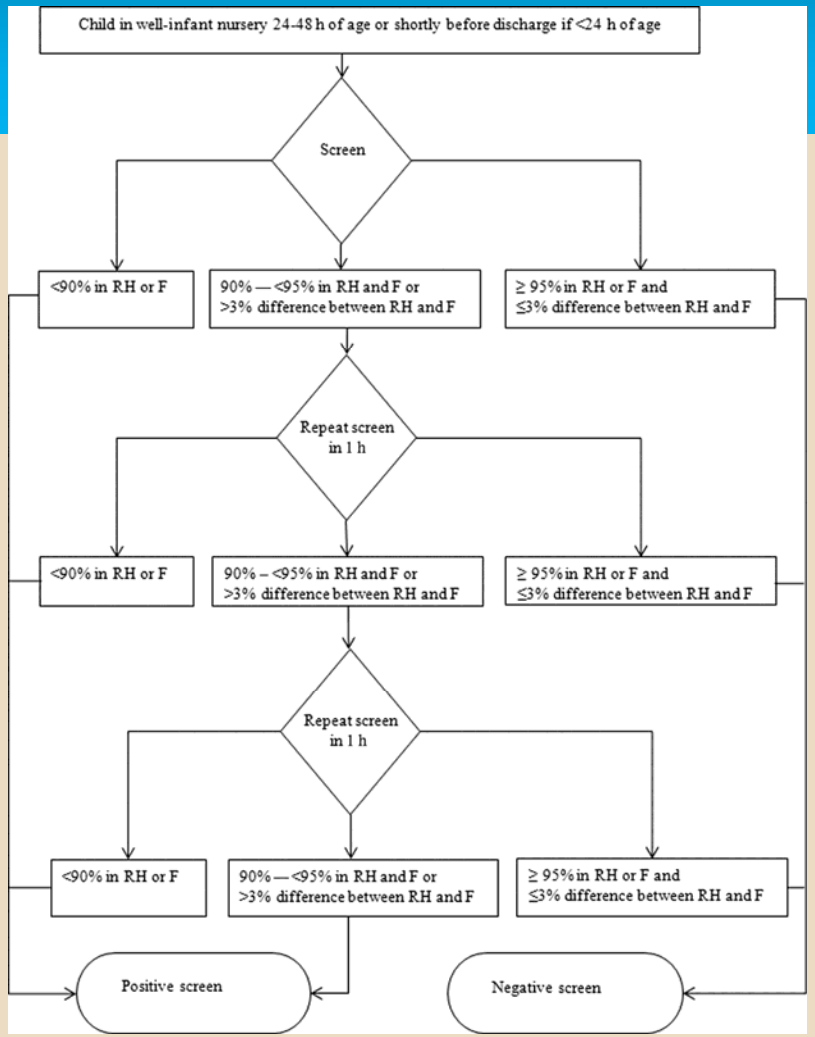
Screening

Baby's First Test video

http://www.youtube.com/watch?feature=player_embedded&v=Lif7kSgHfkw

Interpretation

Pulse-oximetry protocol endorsed by AAP and AHA



Kemper A R et al. Pediatrics 2011;128:e1259-e1267

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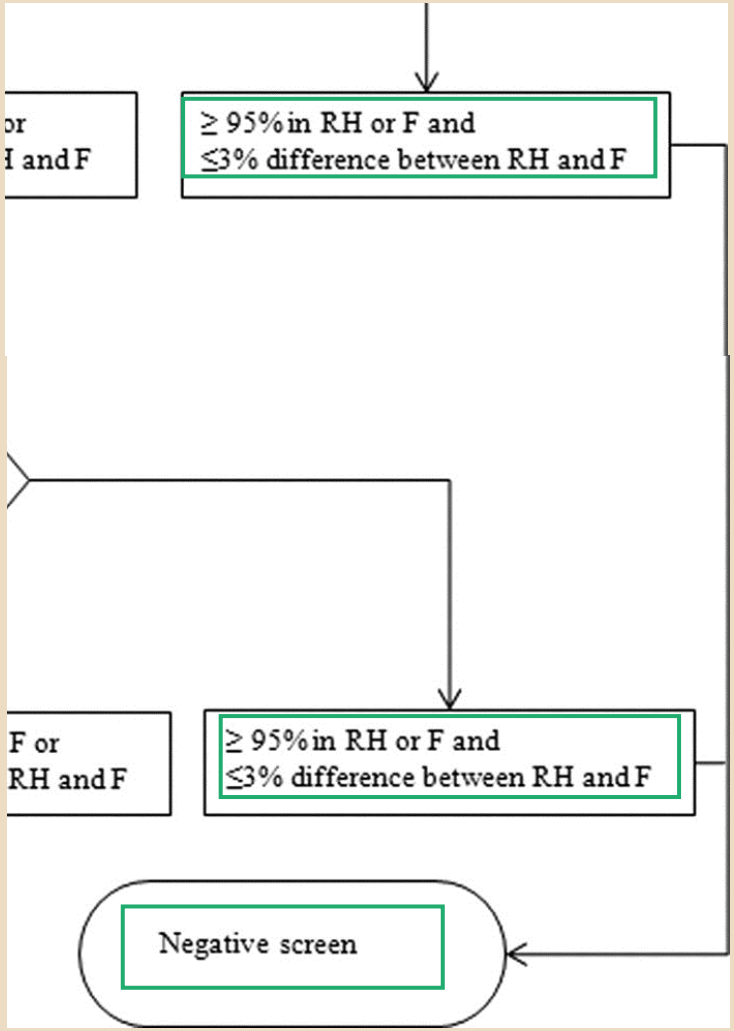
or
I and F
≥ 95% in RH or F and
≤3% difference between RH and F
F or
RH and F
≥ 95% in RH or F and
≤3% difference between RH and F
Negative screen

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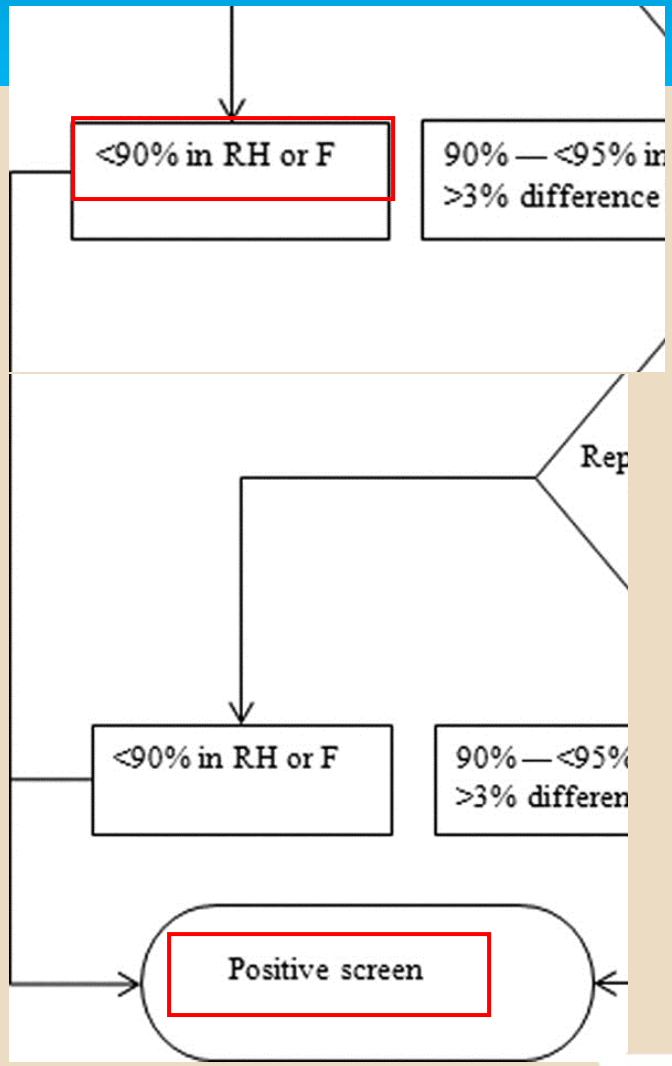
Pulse-oximetry protocol endorsed by AAP and AHA



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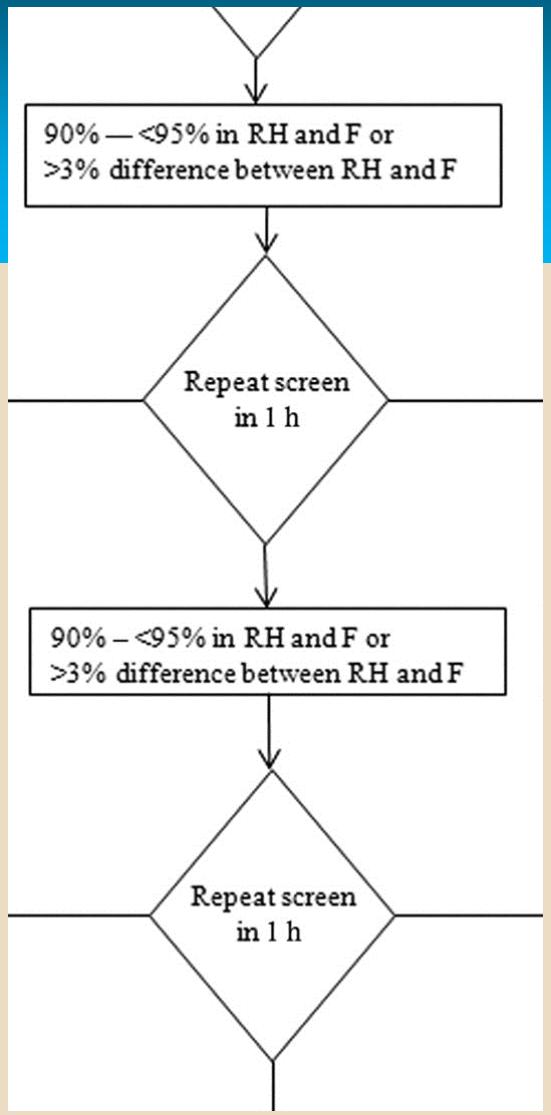
90% — <95% in RH and F or
>3% difference between RH and F
Repeat screen
in 1 h
90% – <95% in RH and F or
>3% difference between RH and F
Repeat screen
in 1 h

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Mueller CCHD Screening Table

Green= Negative Screen (PASS)
Red=Rescreen in 1 hour
Red for 3 consecutive screens= Positive Screen (FAIL)
Red= Automatic Positive Screen (FAIL)

RIGHT HAND	FOOT											<90
100	100	99	98	97	96	95	94	93	92	91	90	*
99	100	99	98	97	96	95	94	93	92	91	90	*
98	100	99	98	97	96	95	94	93	92	91	90	*
97	100	99	98	97	96	95	94	93	92	91	90	*
96	100	99	98	97	96	95	94	93	92	91	90	*
95	100	99	98	97	96	95	94	93	92	91	90	*
94	100	99	98	97	96	95	94	93	92	91	90	*
93	100	99	98	97	96	95	94	93	92	91	90	*
92	100	99	98	97	96	95	94	93	92	91	90	*
91	100	99	98	97	96	95	94	93	92	91	90	*
90	100	99	98	97	96	95	94	93	92	91	90	*
<90	*	*	*	*	*	*	*	*	*	*	*	<90

Positive Screen (FAIL)

- ANY Oxygen Saturation <90% (the **RED ***)
- O2 Saturation 90-94% in both extremities on 3 measures, each separated by 1 hour
- ≥ 95% in either extremity with >3% difference in oxygen saturation between the upper and lower extremity (the **RED** Section of the Table) on 3 measures, each separated by 1 hour

Negative Screen (PASS)

- ≥95% in either extremity with ≤3% absolute difference in oxygen saturation between the upper and lower extremity

- The **Green** Section of the Table

Screening

Individual hospital protocol development:

- Who will screen and when? (remember potential need for evaluation)
- Who will be notified of abnormal screens?
- How will a baby be evaluated after an abnormal screen?
- Who will enter data?
- If a baby is discharged without screening, what will you do?

Evaluation

- Congenital Heart Disease (CHD)
  - Inherited structural abnormality of the heart that is present at the time of birth

- Critical Congenital Heart Disease (CCHD)
  - Any CHD that relies on patency of the ductus arteriosus to provide blood flow to the lungs or to the body (and/or)
  - Any CHD that requires surgical or catheter based intervention during the first month of life

Evaluation

- Cyanotic CHD
  - Any CHD associated with oxygen saturation less than 95%
  - Due to mixing of oxygen “rich” blood from the lungs and oxygen “poor” blood from the body

- Some forms of CCHD are associated with cyanosis (i.e. oxygen saturation less than 95%) and are considered cyanotic critical congenital heart disease (CCCHD)

Evaluation

- Pulse oximetry is a screening tool used to help identify babies with cyanotic CHD

- Pulse oximetry may also help detect babies with other forms of CHD or other non- heart related conditions (i.e. infection, lung disease)

Evaluation

- Cyanotic CHD that may be identified by pulse oximetry screening include (not limited to):
  - Hypoplastic Left Heart syndrome (HLHS)
  - Pulmonary atresia (PA)
  - Tetralogy of Fallot (TOF)
  - Total anomalous pulmonary venous return (TAPVR)
  - Dextro-Transposition of the Great Arteries (dTGA)
  - Tricuspid atresia (TA)
  - Truncus arteriosus

Evaluation

- All children require a detailed physical examination in the nursery or neonatal intensive care unit

- Children with “positive” pulse oximetry screening test results (i.e. sats < 95% or greater than 3% difference in saturation between upper and lower extremity) <u>require</u> urgent clinical evaluation and may undergo an echocardiogram, interpreted by a pediatric cardiologist; consultation with a pediatric cardiologist may be required

Evaluation

- In the event that an echocardiogram cannot be performed, children with concern for CCHD may require transfer to another medical center

- In cases of suspected CCHD, a diagnosis should be made prior to discharge to home

Evaluation

- Families should be advised that an out-of-range screening result does “not necessarily mean that your baby has CCHD”
- Families should be informed that pulse oximetry <u>does not detect all forms of CHD</u>
- Pediatricians and primary providers should be informed of results

Evaluation

- Families should be informed of the following:

  <u>Warning signs of CHD</u>

  - Poor feeding
  - Poor urine output (less than 3 solid wet diapers per day)
  - Tires easily or sleeps a lot
  - Poor weight gain
  - Often irritable or difficult to console
  - Pale, bluish color

Surveillance

- DHMH will provide surveillance of screening.
- Data will be entered into OZ for this function.
- Provide Debra Harper-Hill, RN with contacts needing access

Surveillance

- Nurse will contact providers regarding abnormal screens to determine the evaluation process and ultimate diagnosis for the infant..
- To evaluate efficacy of protocol and challenges to implementation.
- Quarterly feedback to birthing facilities regarding numbers screened and false positives.

Contact Information

- Webpage: http://fha.dhmh.maryland.gov/genetics/SitePages/CCHDScreeningProgram.aspx
- Debra Harper-Hill, RN 410-767-6625 debra.harperhill@maryland.gov
- Debbie Badawi, MD 410-767-5592 deborah.badawi@maryland.gov

Maryland Prevention and Health Promotion Administration

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