

Pulse Oximetry Screening for Critical Congenital Heart Disease



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Universal newborn screening all 50 states



Newborn hearing screen mandated in 44 states



2009 American Academy Ped

- Data not strong enough
- Many false positives
- 2010 Congress authorized Health and Human Services Secretary's Advisory Committee on Heritable Disorders in Newborns and Children (SACHDNC) to provide guidance to the Sec of HHS about conditions to be included in newborn screening.



Maryland First State to Mandate Newborn Pulse Oximetry screening June 2011



- <http://www.cbsnews.com/video/watch/?id=7377745n&tag=mg;eveningnews>



National Recommendation

- September 2011 Kathleen Sebelius, Sec of Health and Human Services (HHS) recommended Universal Pulse Oximetry Screening
- Recommended Uniform screening panel (RUSP).
- November 2011 AAP Strategies of Implementing Screening for Critical Congenital Heart Disease.
- January 2012. American Academy of Pediatrics endorsed the HHS Secretary's recommendation



Dylan Gordon and NJ Gov. Chris Christie



NJ Law enacted 8/31/11 and Dylan identified 9/1/11

Photo: Office of the Governor, state of NJ.

MultiCare – Tacoma, WA

Did you know...

- Many cases of sudden cardiac death in teens and young athletes are caused by undiagnosed heart defects.
- Adults with CHDs require lifelong medical care from trained heart specialists; follow-up visits are recommended from every six months to every five years, depending on the type of defect.
- Women with heart defects should check with their cardiologist before becoming pregnant. Women with CHDs, or a family history of heart defects, may need careful monitoring by a high-risk obstetrician, as well as their cardiologist, throughout pregnancy.

PARTICIPATING PEDIATRIC CARDIOLOGY PRACTICES:

NorthWest Children's Heart Care

253.396.4868

Northwest Pediatric Heart Specialists

253.272.1812

Swedish Pediatric Cardiology

206.215.2700

This publication sponsored in part by the Fraternal Order of Eagles.

WAS THIS HELPFUL?

- Did you learn something new?
- Did you recognize the signs of CHD in a child that was then diagnosed or treated for a heart defect?

If so, we would like to hear from you. Please call or write to us and share your story.

FOR MORE INFORMATION

For questions or comments, please contact
Mary Bridge Pediatric Heart Center

Newborn Screening for Heart Defects

Information for Parents



multicare.org

Allenmore Hospital
Good Samaritan Hospital

Deaconess RCPs started the process March 6, 2012

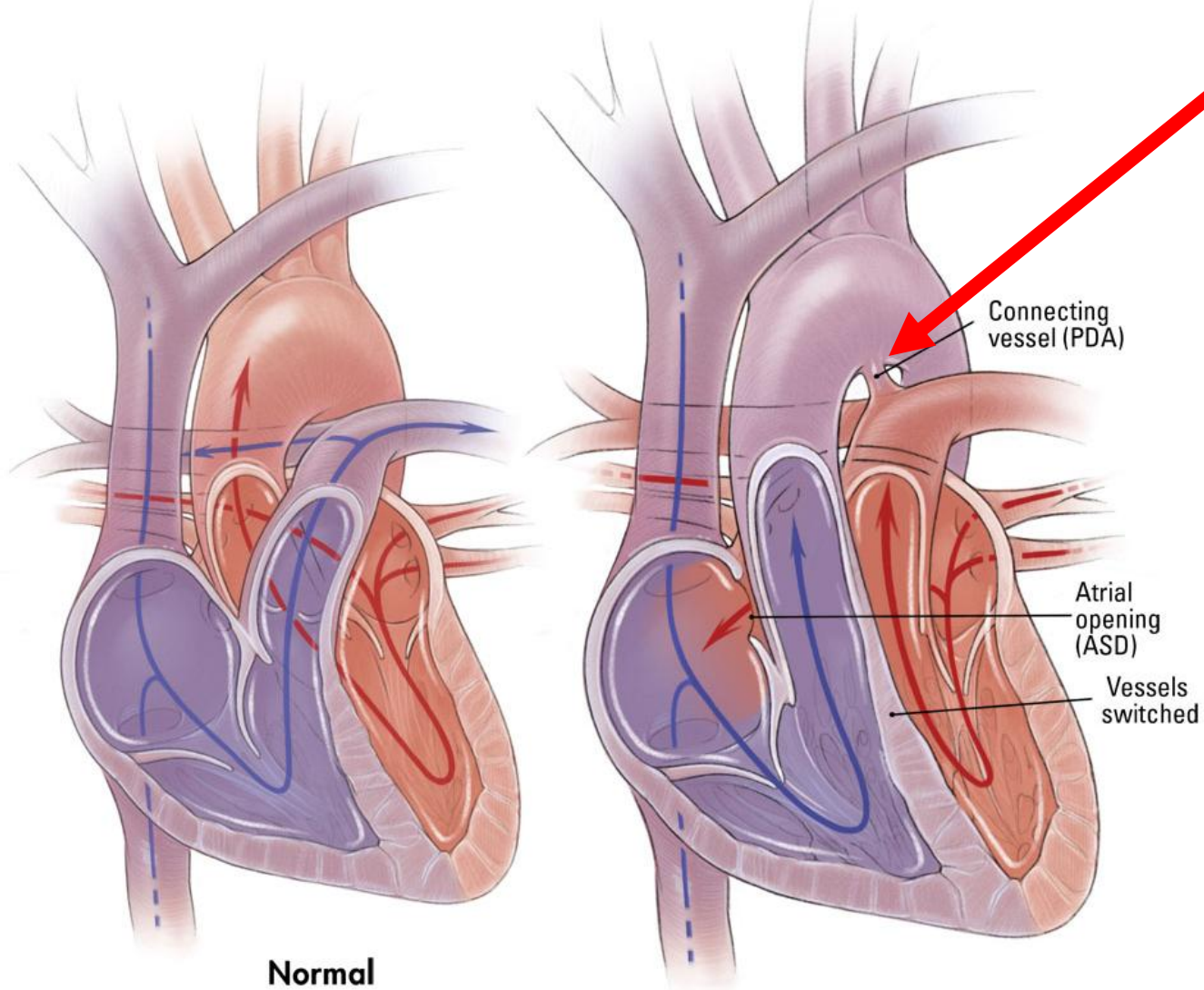


Why do Pulse Ox Screening?

- Cardiac malformations are the most common congenital defects.
- Prevalence 6-21/1000 live births.
- 1-1.8/1000 live births have duct dependent malformations (deWahl)
- Of those, 25% are critical congenital defects
- In US, 24% of infant deaths are due to CHDefects
- Risk of mortality and morbidity decreases with identification in Newborn period



Ductal Dependent Transposition



Normal

Transposition of great arteries

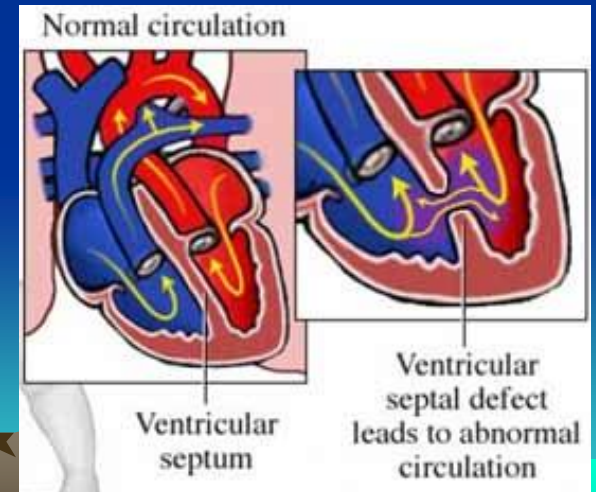
Are all heart defects detected
because you hear a murmur?



Are all heart defects detected because you hear a murmur?

- May be absent or misleading
- Heart murmurs occur in non-critical heart disease
- Typically diagnosed later in life
- Larger the “shunt,” less resistance

• www.empowerher.com



Can we see CCHD on a prenatal ultrasound?

Children's Hospital Boston



Can we see CCHD on a prenatal ultrasound?

- Yes, however
- Less than 1/2 of CCHD are picked up on prenatal ultrasound



Pulse Oximetry Screening for Critical Congenital Heart Disease

- Purpose
- Early detection of CCHD
 - Prevent childhood deaths
 - Prevent childhood injury
- Avoid late detection (circulatory collapse)
- Without screening, risk of leaving hospital with undiagnosed ductal dependent defect is 30%



Pulse Oximetry Screening for Critical Congenital Heart Disease

- Sweden 40,000 newborns screened with *new generation* pulse oximetry
- mortality among infants discharged with an undiagnosed critical heart defect was 18%
- Deaths for those diagnosed before leaving hospital - 0.9%.
- False positive = 0.17%

• Sahlgrenska Academy U of Gothenburg, Sweden deWahl Granelli et al 2010

• Princess Estelle of Sweden 2/3/12



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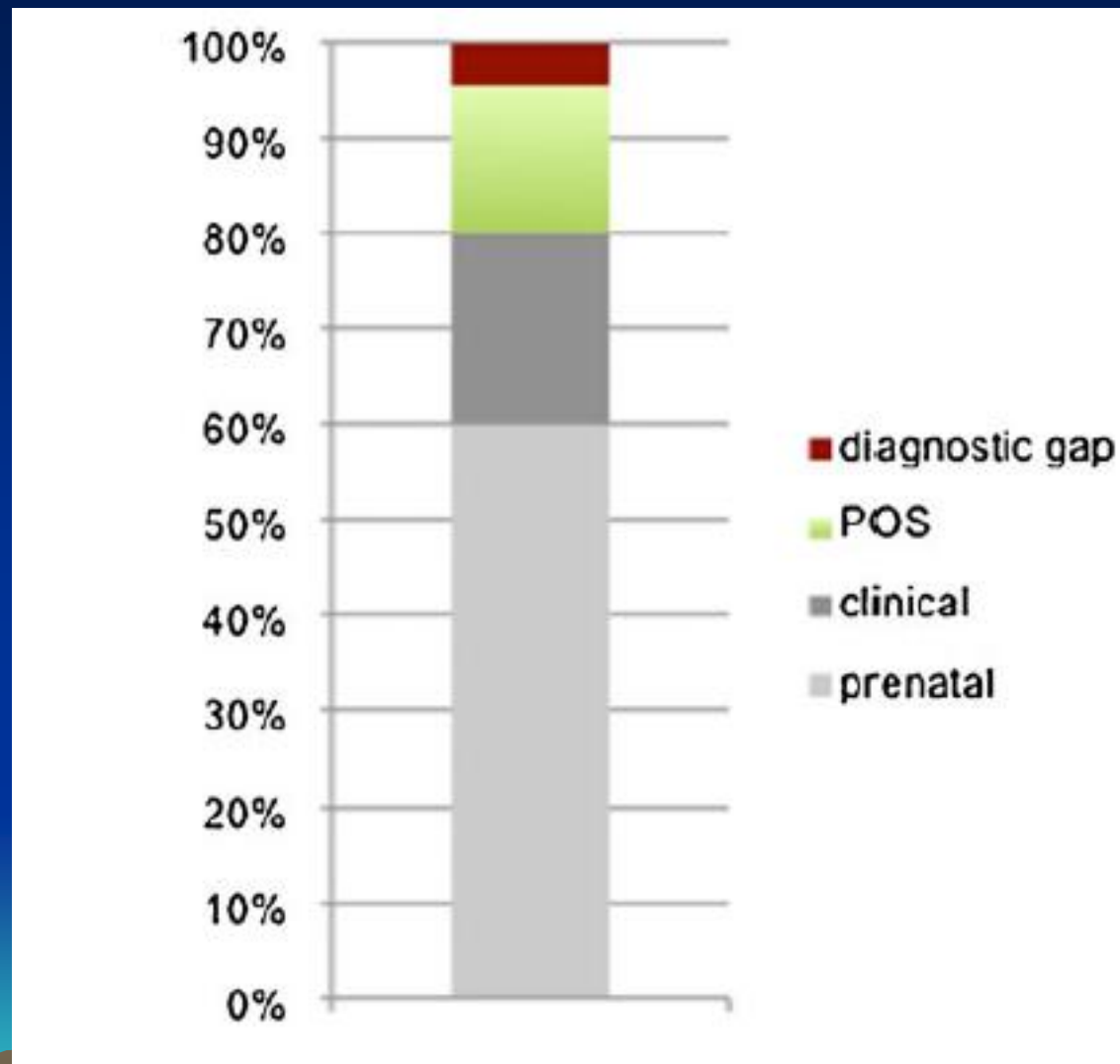


Pulse Oximetry Screening for Critical Congenital Heart Disease

- 41,445 newborns Germany ≥ 37 wks
- Pulse Oximetry Screening (POS)
 - true positive in 14
 - False positive in 40 (0.10%)
 - Healthy = 12
 - PPHN = 15
 - Sepsis = 13
 - False negative 4 (3 were studied too early 4,6 and 10 hours age and PO were 94-96%)
- Riede, FT Wörner, C et al (2010). Eur J Ped

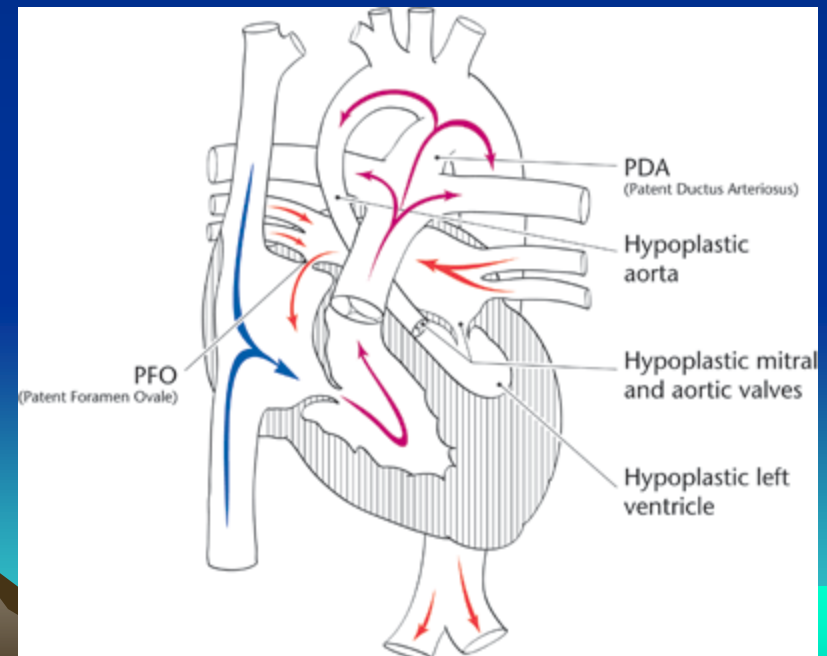


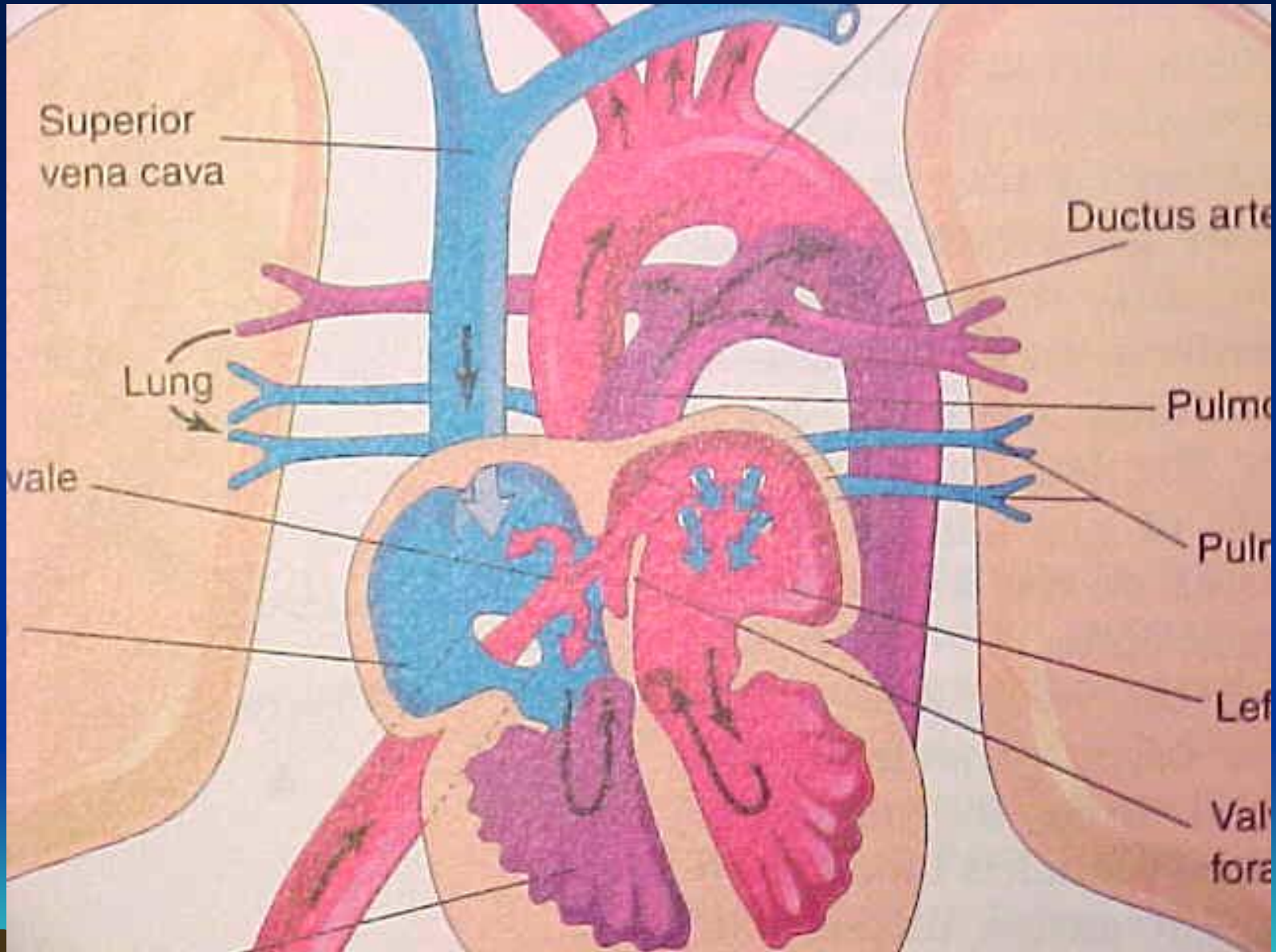
Timing and Method of diagnosis cCHD



Timing of symptoms depends upon

- The actual defect
- Changes in pulmonary vascular resistance and
- Changes in systemic vascular resistance
- Closure of the ductus



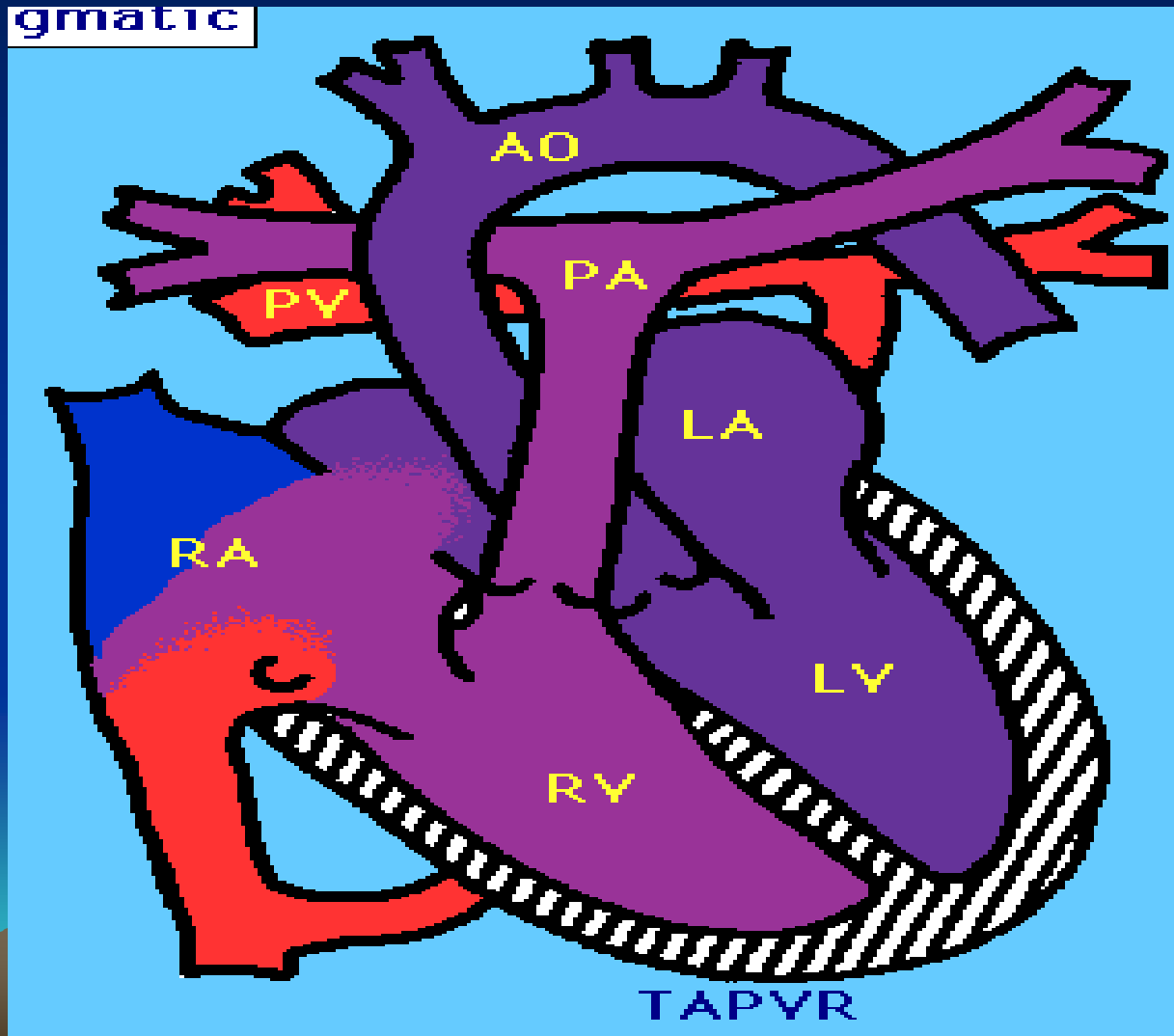


Pulse oximetry \approx 24 hours of age

- Seven Critical Congenital Heart Defects (CCHD) potentially can be detected among some babies using pulse oximetry screening
- Other heart defects can be just as severe as these seven CCHDs and also require treatment soon after birth.
- However, pulse oximetry screening **may not** detect these heart defects as consistently as the seven disorders listed as CCHDs



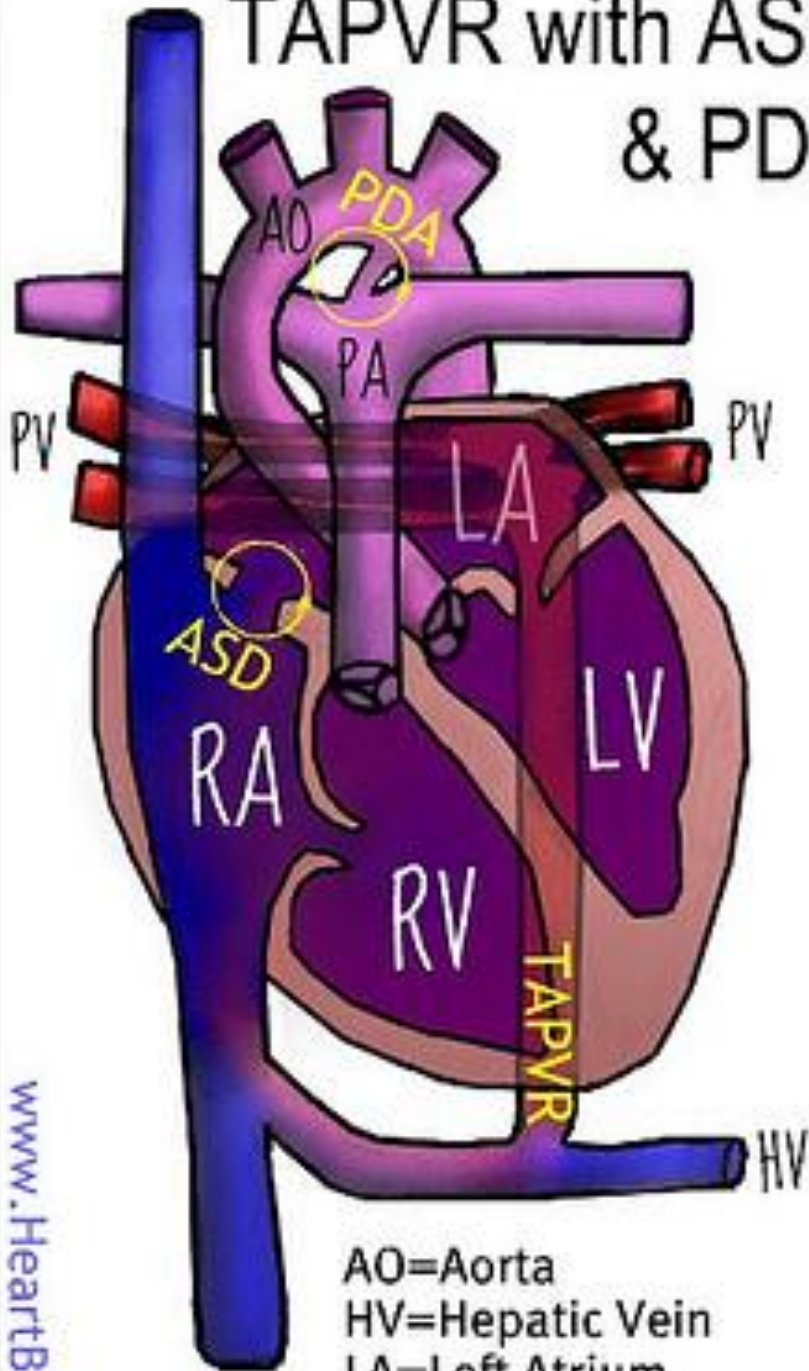
Obstructed Total Anomalous Venous Return



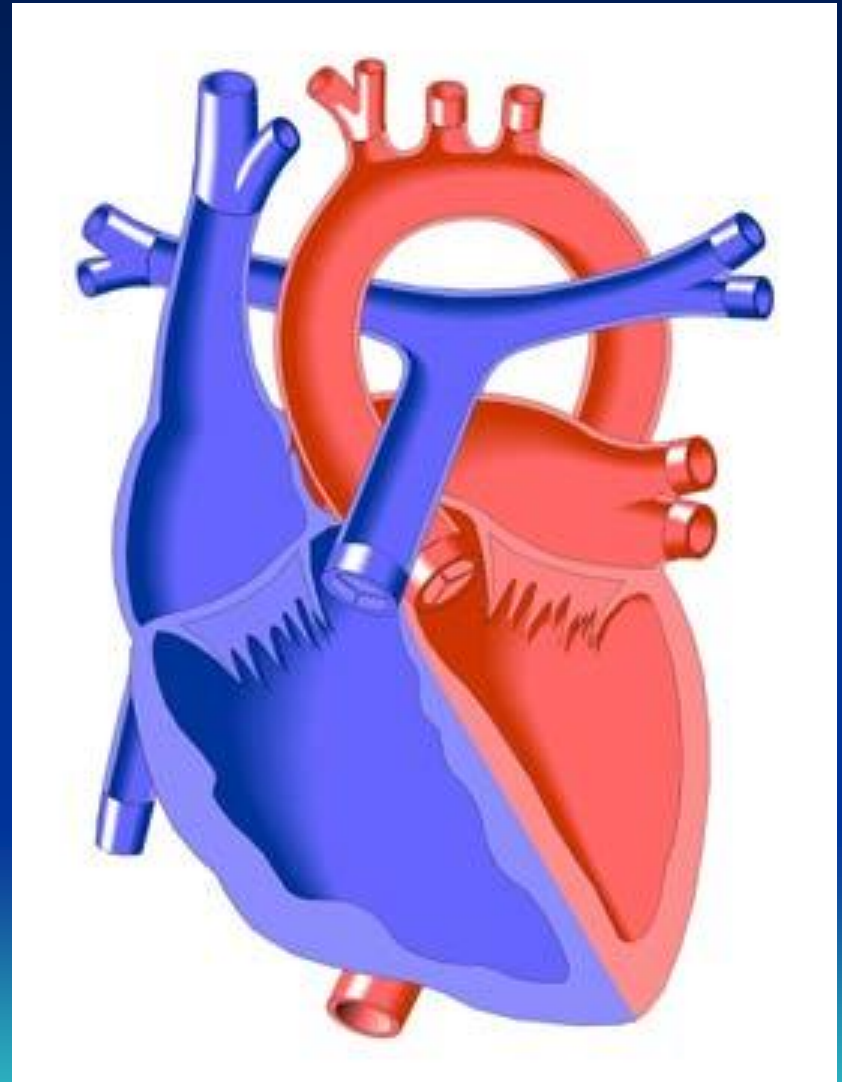
http://www.youtube.com/watch?v=AixPDa3hYik&feature=results_video&playnext=1&list=PL0A12D0B4DD5663E5



TAPVR with ASD & PDA

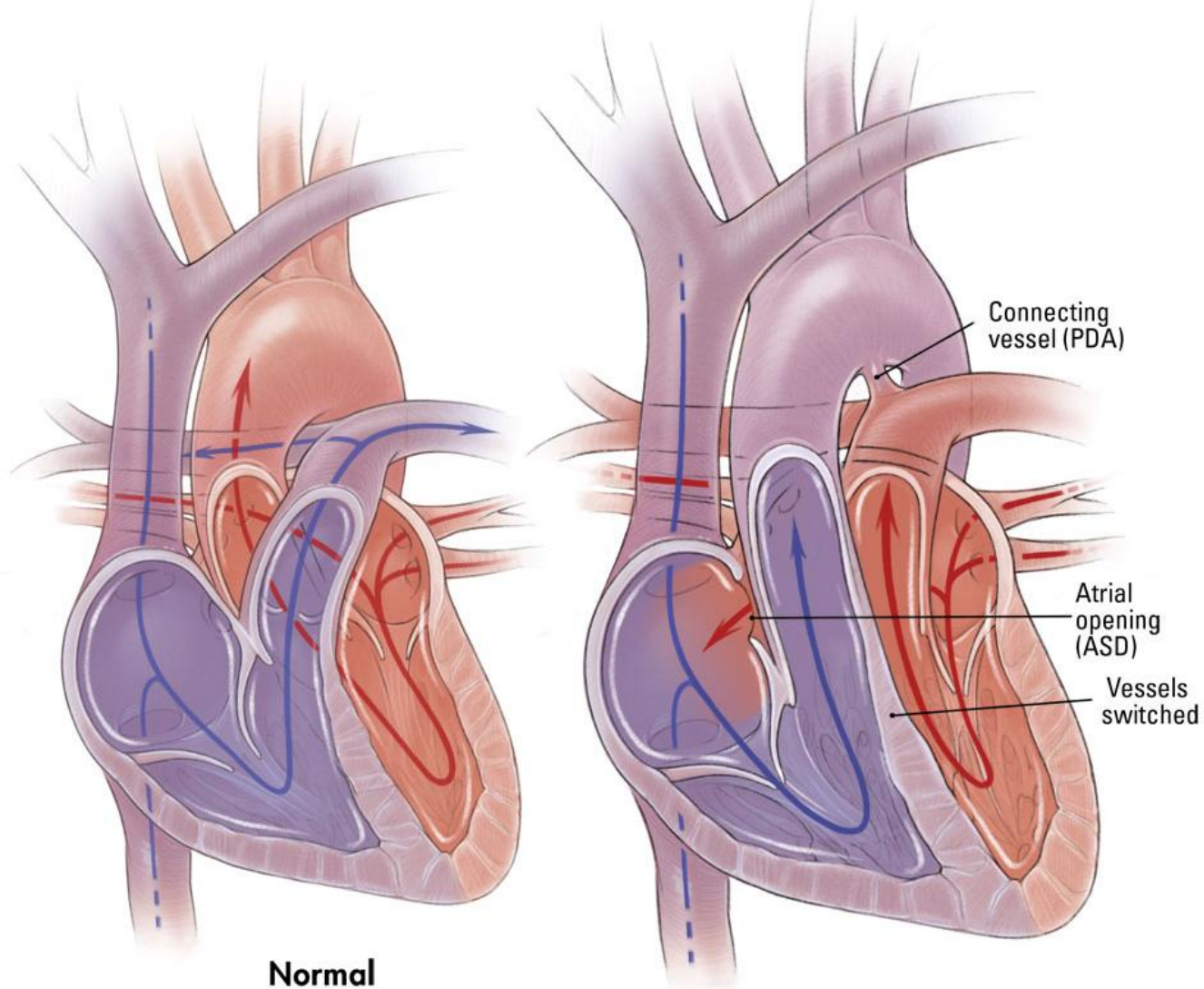


AO=Aorta
HV=Hepatic Vein
LA=Left Atrium



Timing of Symptoms

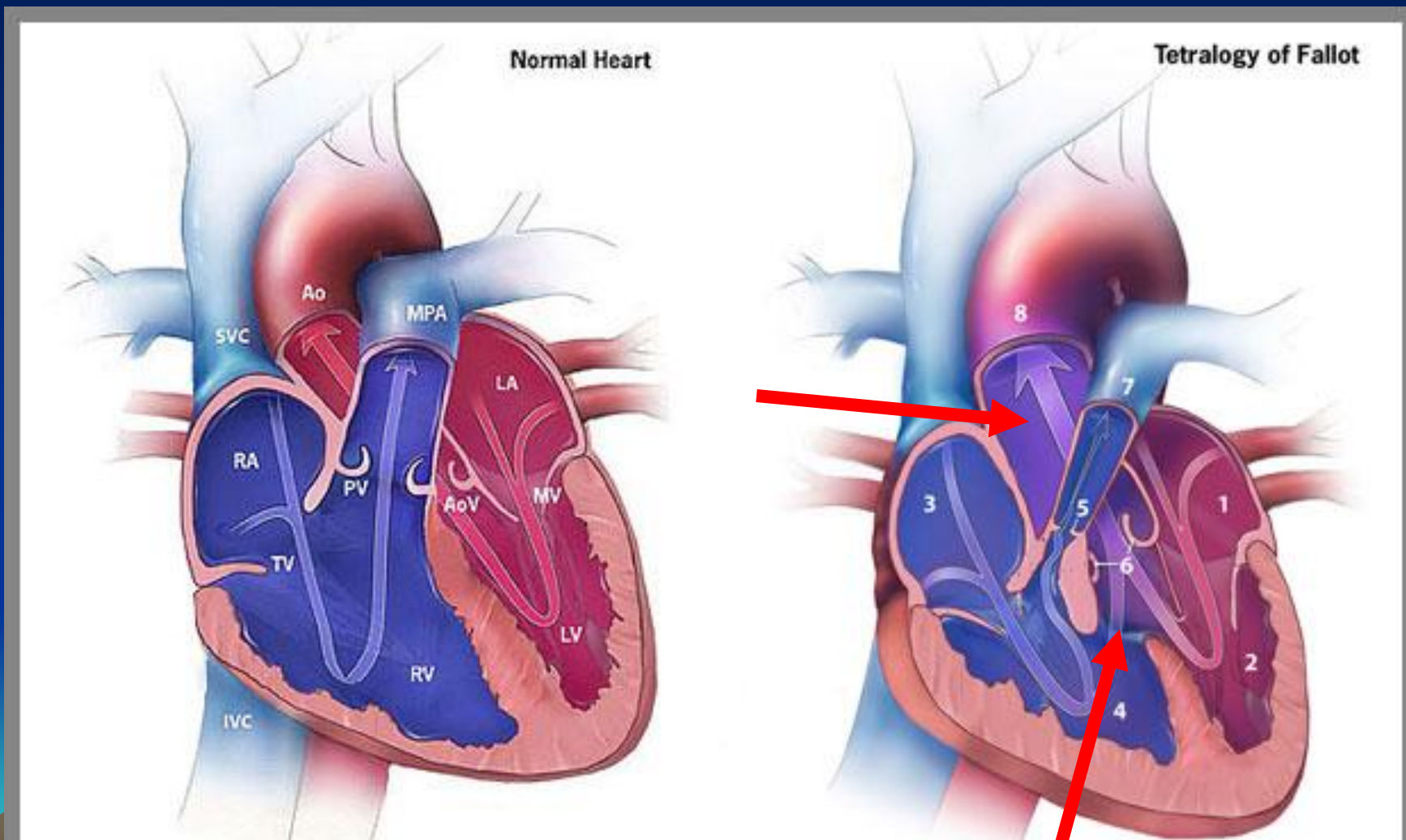
Hours after birth



Normal

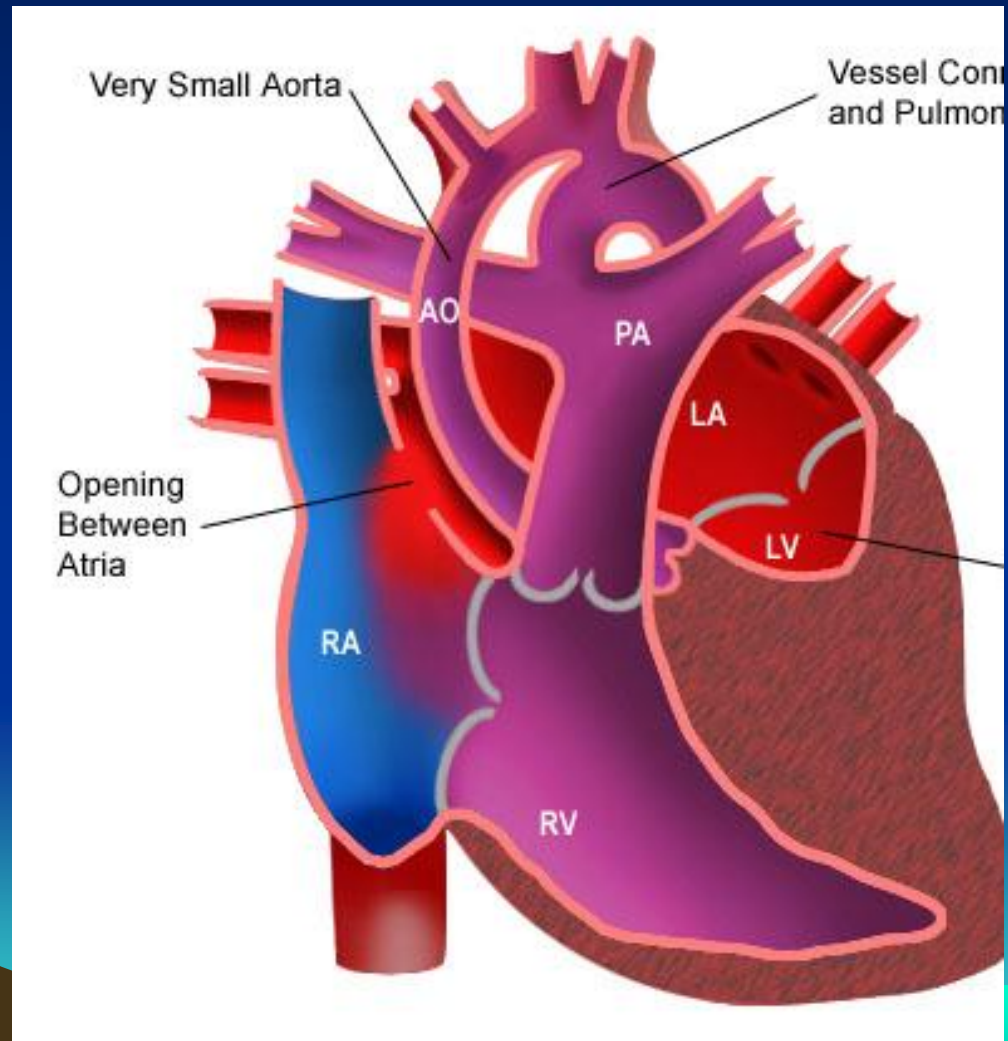
Transposition of great arteries

Obstructed Right Side Tetralogy of Fallot

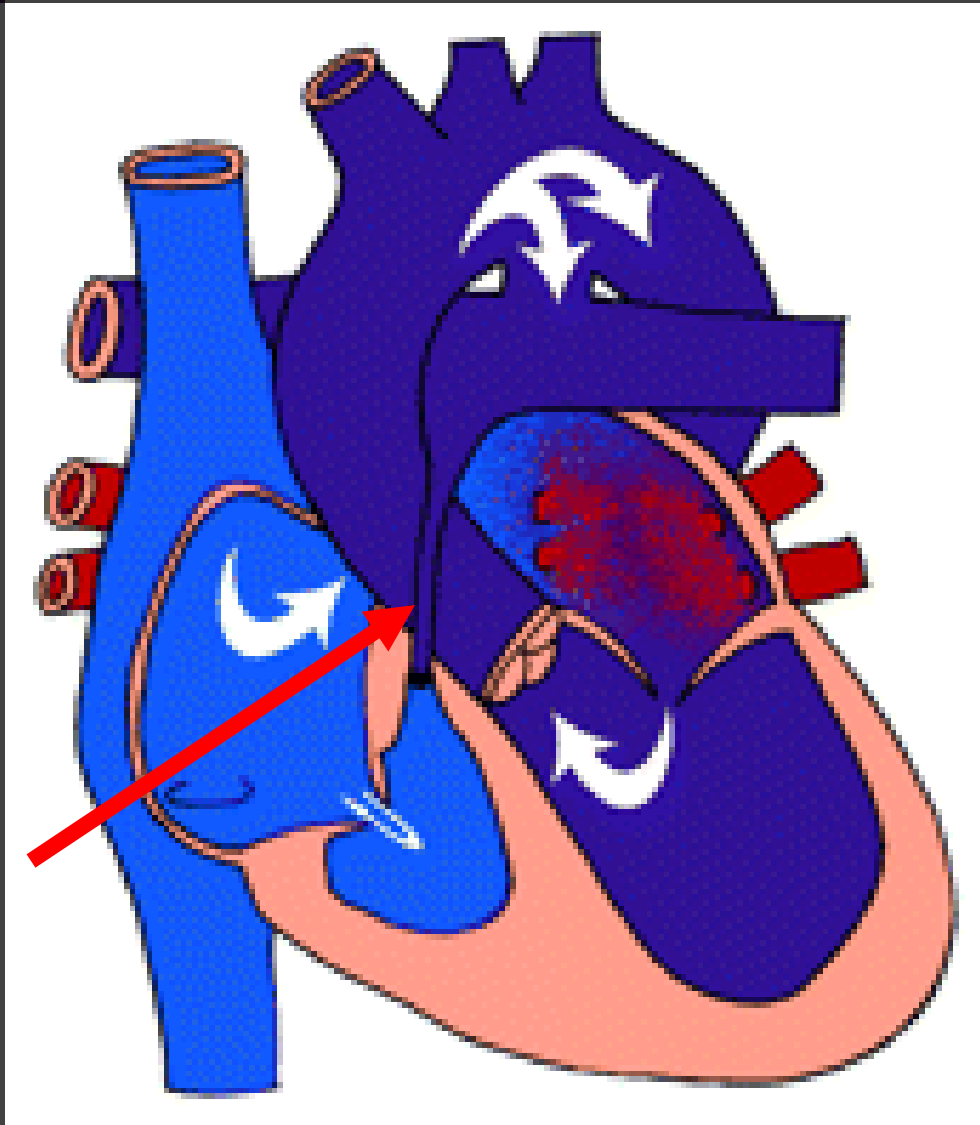


Obstructed L side

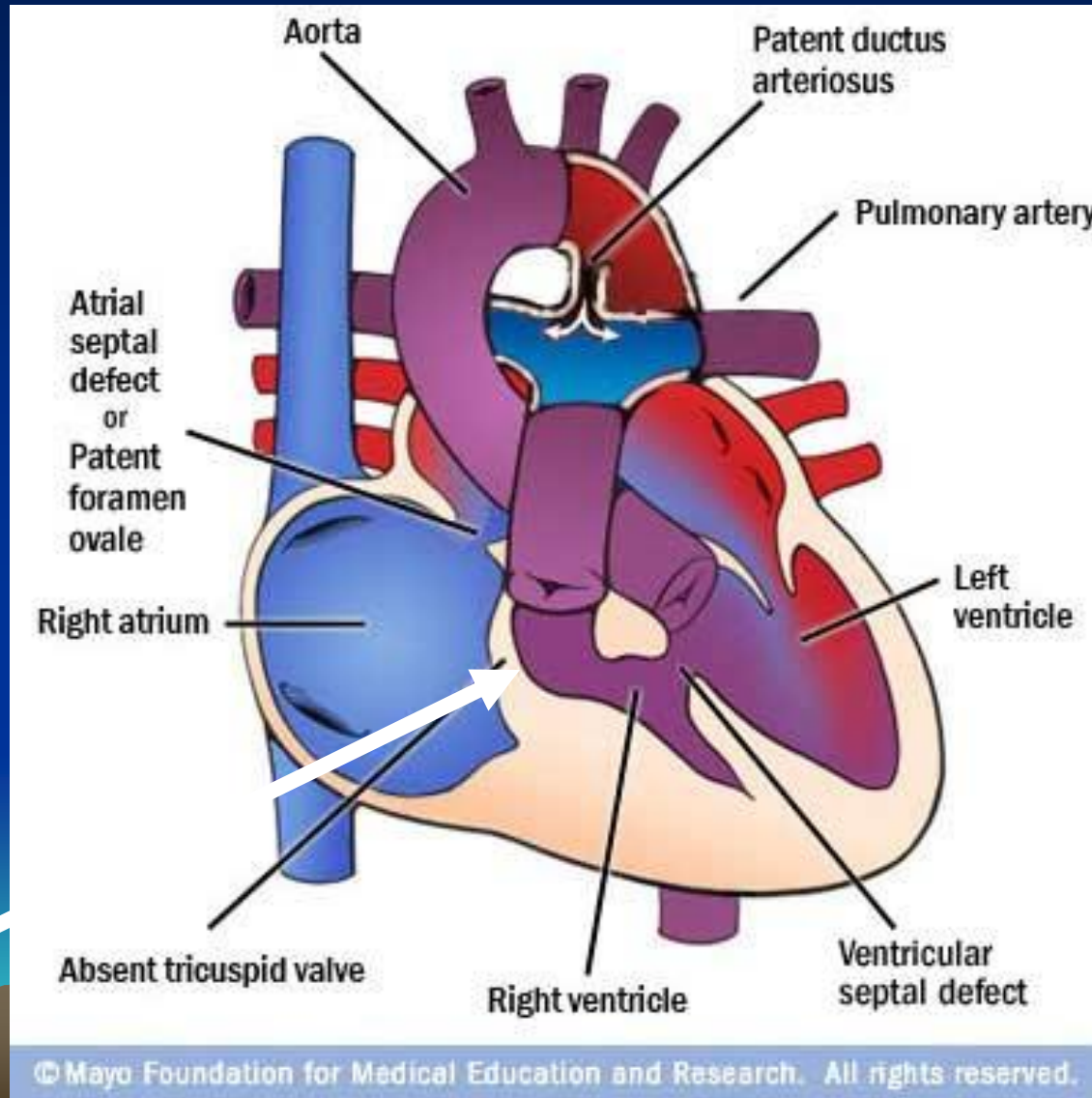
Hypoplastic Left Ventricle



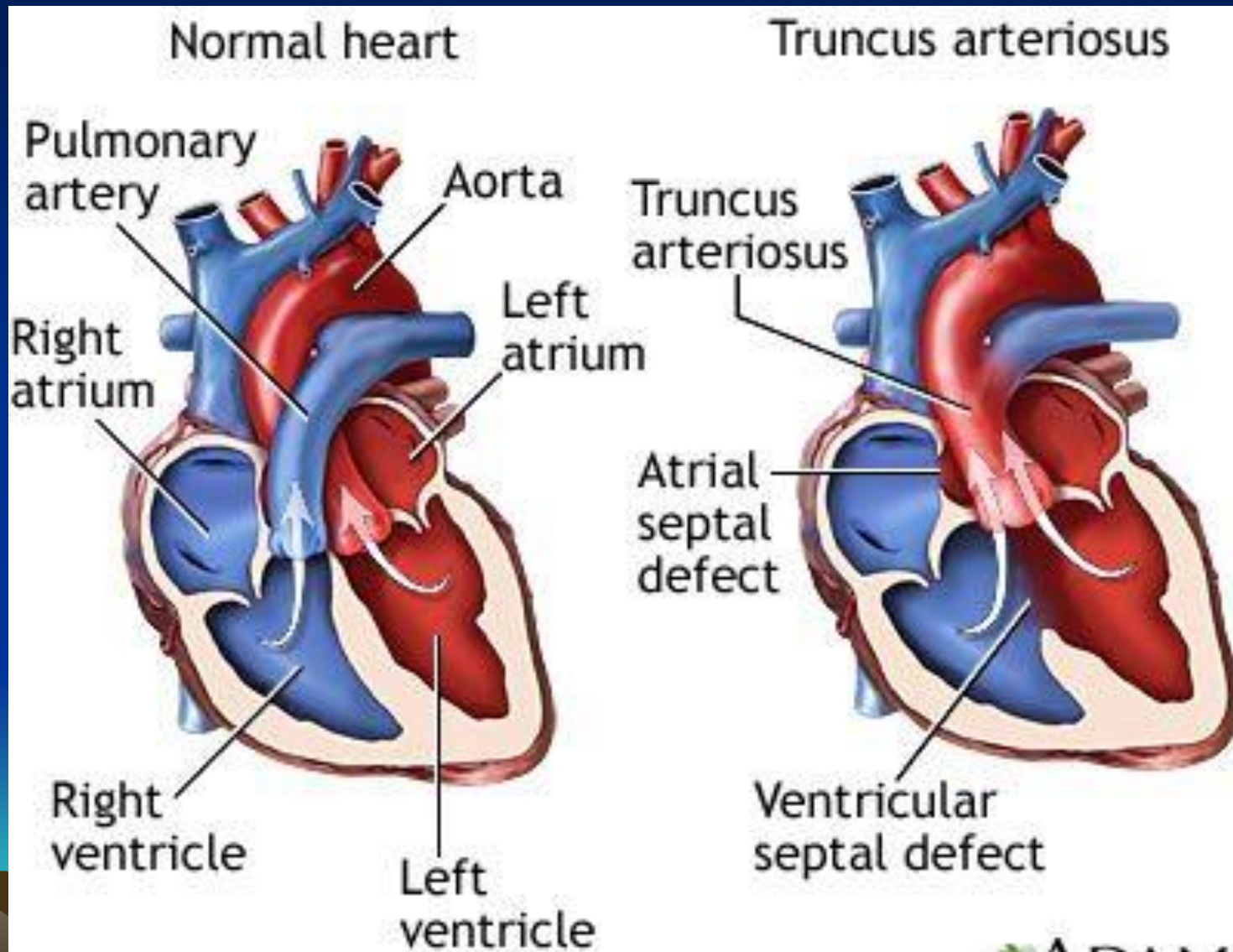
Pulmonary atresia intact septum



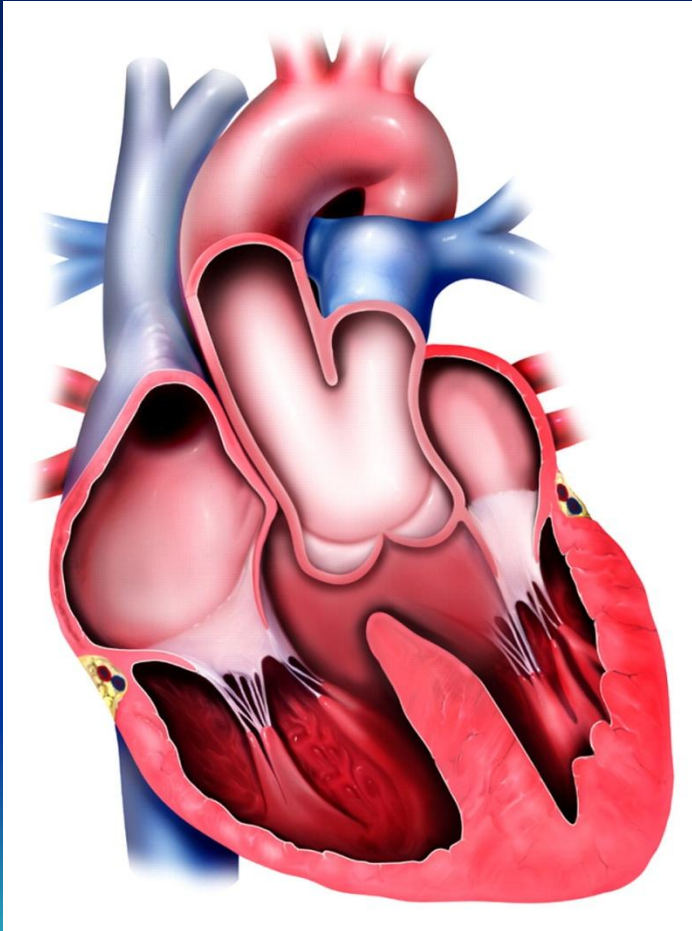
Tricuspid Atresia



Truncus Arteriosus



Truncus Arteriosus (One main trunk)



Gaca A M et al. Radiology 2008;248:44-60

Parent information from CDC

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

Pulse Oximetry Screening for Critical Congenital Heart Defects

- Babies with a critical congenital heart defect (CCHD) are at significant risk for death or disability if their condition is not diagnosed soon after birth.
- Pulse oximetry newborn screening can identify some infants with a CCHD before they show signs of the condition.
- Once identified, babies with a CCHD can be seen by cardiologists and can receive special care and treatment that can prevent death or disability early in life.
- Certain hospitals routinely screen all newborns using pulse

Understanding Critical Congenital Heart Defects

- **Congenital heart defects (CHDs) account for 24% of infant deaths due to birth defects.**
- **In the United States, about 4,800 (or 11.6 per 10,000) babies born every year have one of seven *critical congenital heart defects* (CCHDs, which also are known collectively in some instances as *critical congenital heart disease*).**
- **These seven CCHDs are:**
 - » **Hypoplastic left heart syndrome**
 - » **Pulmonary atresia (with intact septum)**
 - » **Tetralogy of Fallot**
 - » **Total anomalous pulmonary venous return**
 - » **Transposition of the great arteries**
 - » **Tricuspid atresia**



How long does it take and how much does it cost?

- Each screen takes from 1 to 3 minutes.
- Cost: \$1.20 to \$10.00 per baby

- AAP approached AMA to develop payment code for reimbursement.



Who will receive screening

- All healthy newborn infants in the newborn nursery
- As close to discharge as possible if < 24 hours of age



When should the screening be done?

- Aim for at least 24 hours of age or older
 - Functional ductus arteriosis closure

This reduces the number of false-positive results.



How is it to be done?

- Place an oximeter on the baby's **RIGHT** hand/wrist and a foot.
- May be done sequentially or concomittantly



CCHD detection is increased when oximetry devices are

- Motion Tolerant
- and
- Low perfusion capable



PASS

15



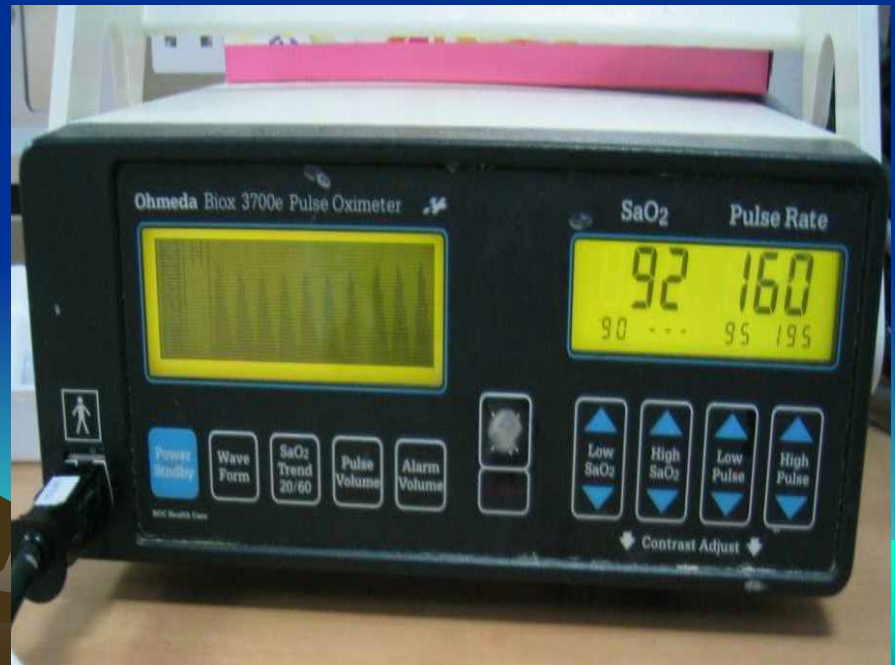
Pass

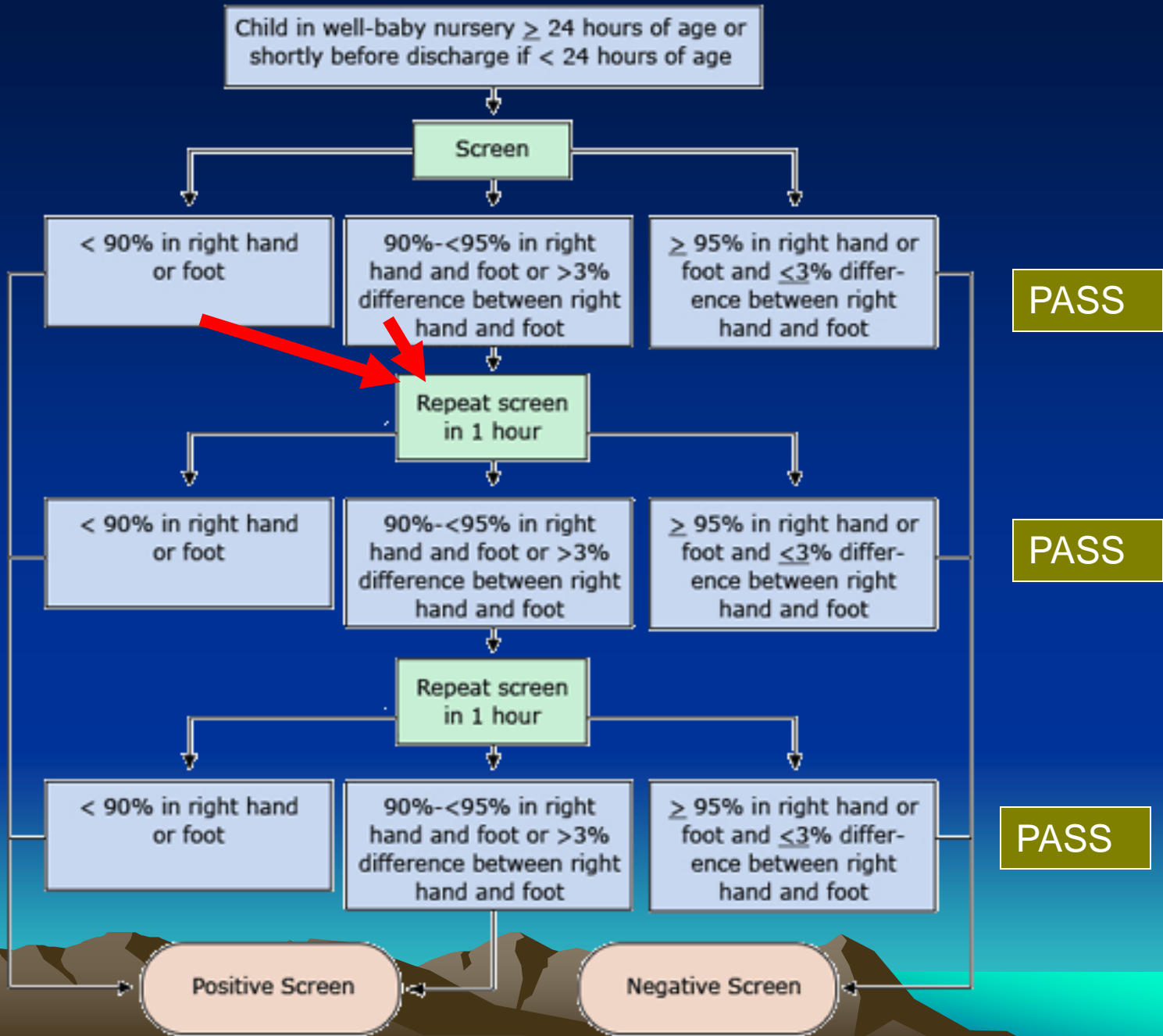
- Anytime the saturation is $\geq 95\%$ in either extremity
- AND
- $\leq 3\%$ absolute difference between upper and lower extremity.
- Considered a pass



ReDo

94 or Less





Child in well-baby nursery ≥ 24 hours of age or shortly before discharge if < 24 hours of age

Screen

$< 90\%$ in right hand or foot

90% - $<95\%$ in right hand and foot or $>3\%$ difference between right hand and foot

$\geq 95\%$ in right hand or foot and $\leq 3\%$ difference between right hand and foot

PASS

Repeat screen in 1 hour

$< 90\%$ in right hand or foot

90% - $<95\%$ in right hand and foot or $>3\%$ difference between right hand and foot

$\geq 95\%$ in right hand or foot and $\leq 3\%$ difference between right hand and foot

PASS

Repeat screen in 1 hour

$< 90\%$ in right hand or foot

90% - $<95\%$ in right hand and foot or $>3\%$ difference between right hand and foot

$\geq 95\%$ in right hand or foot and $\leq 3\%$ difference between right hand and foot

PASS

Positive Screen

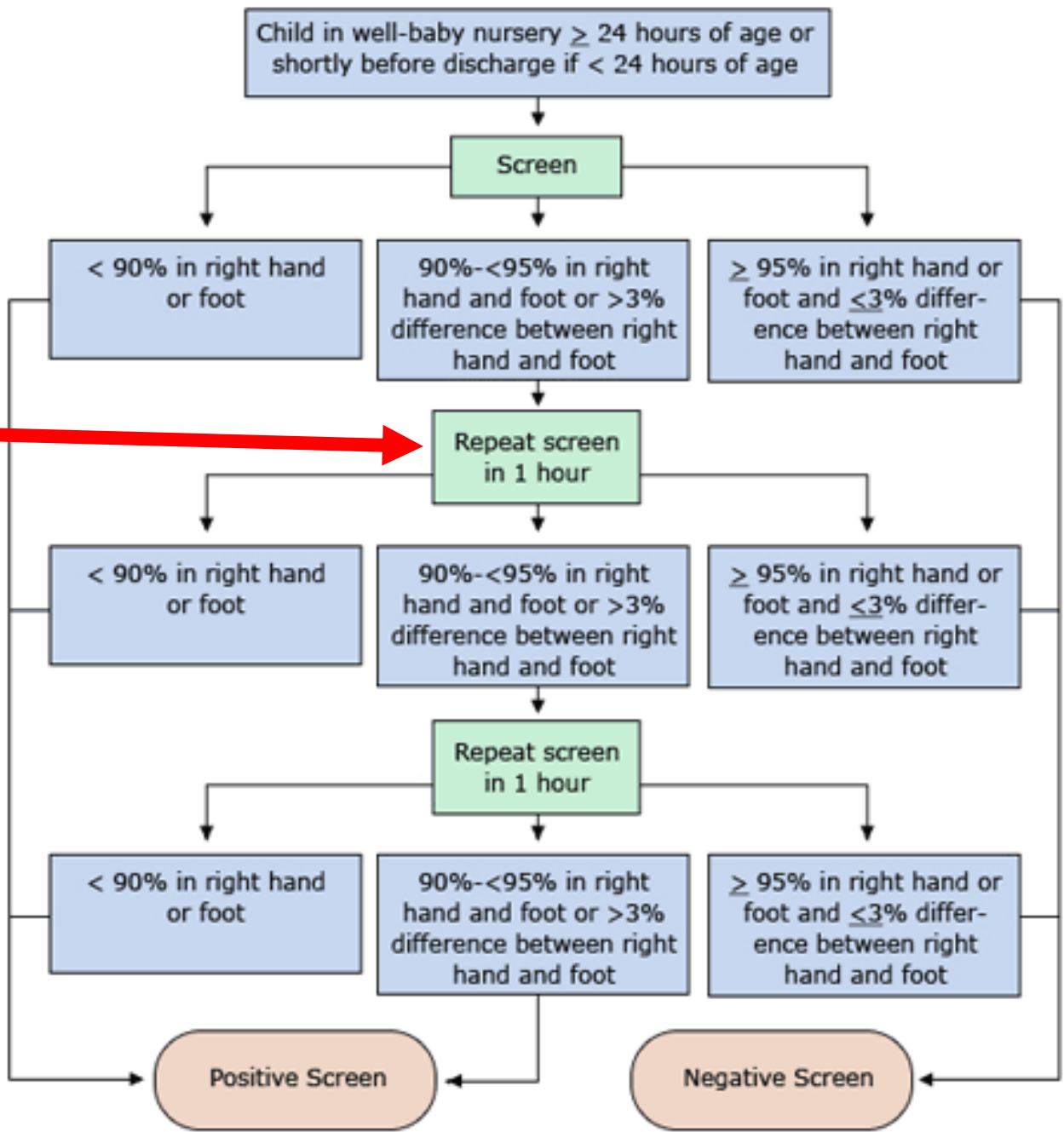
Negative Screen

Repeat oximeter measure screen in one hour when

O₂ sat is 94 or less either Right hand OR
either foot

- Or
- Greater than 3% deviation right hand and
either lower extremity



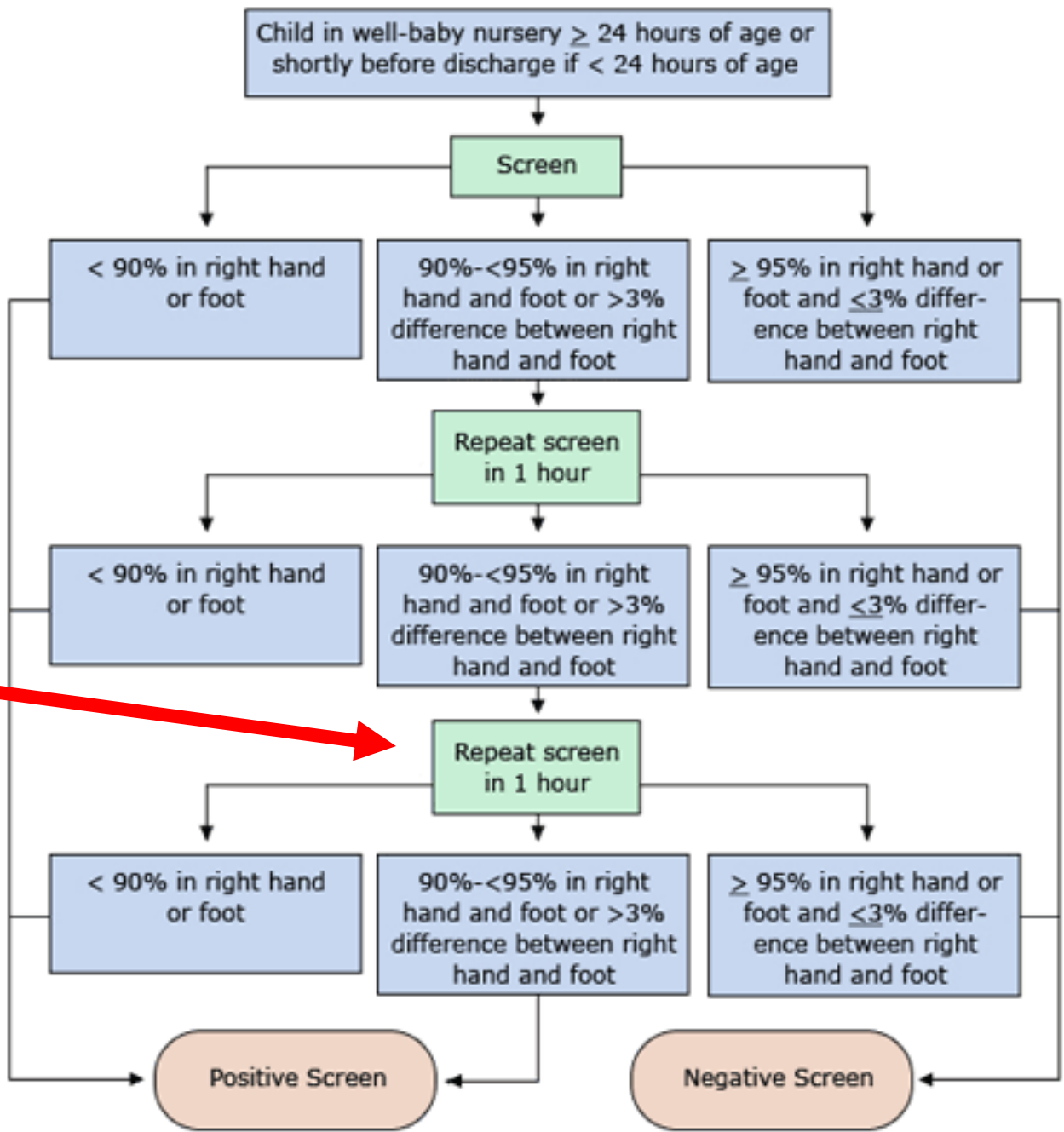


If after 2nd screen, saturations continue to be low, repeat oximeter measure in one hour

O2 sat is 90 – 94%

- Or
- Greater than 3% deviation right hand and either lower extremity





Positive screen after 3rd low or varied measurement

- Now what do we do?

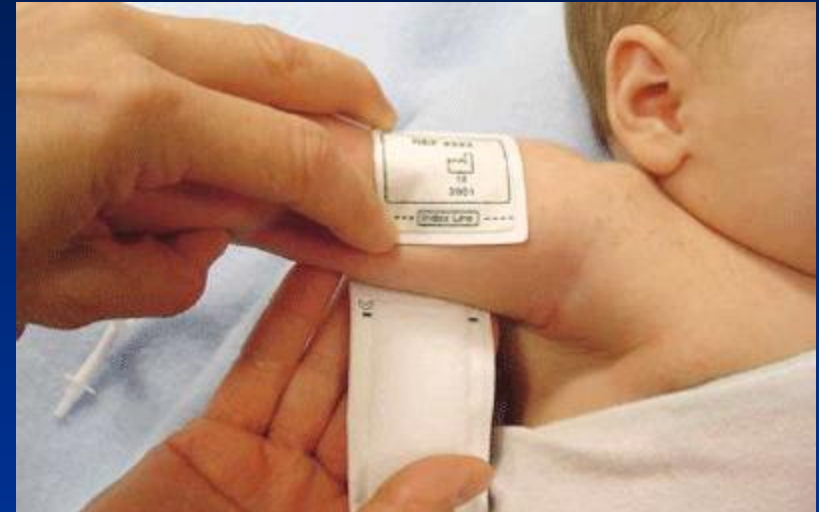
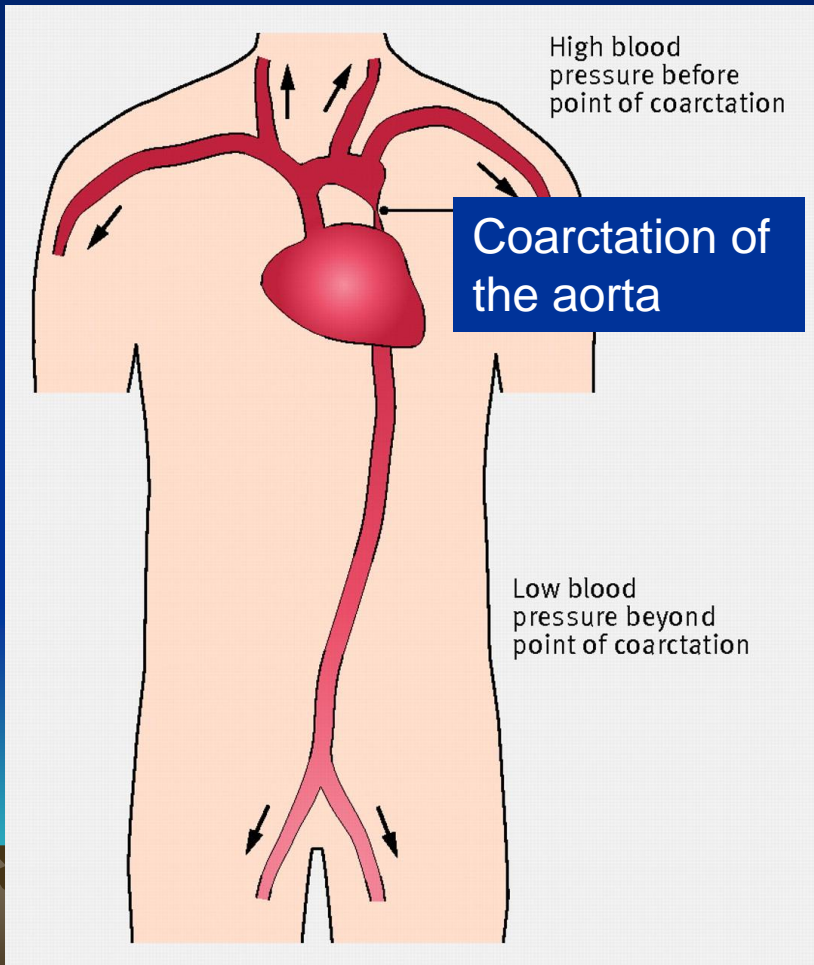


Physical examination



- Saturations that do not meet criteria.
- Notify Primary Care Provider
- Exam by licensed HCP
- Echocardiogram read by a Peds cardiologist

Upper and Lower BP



What happens when there is no pediatric cardiologist in a community?



Echocardiogram

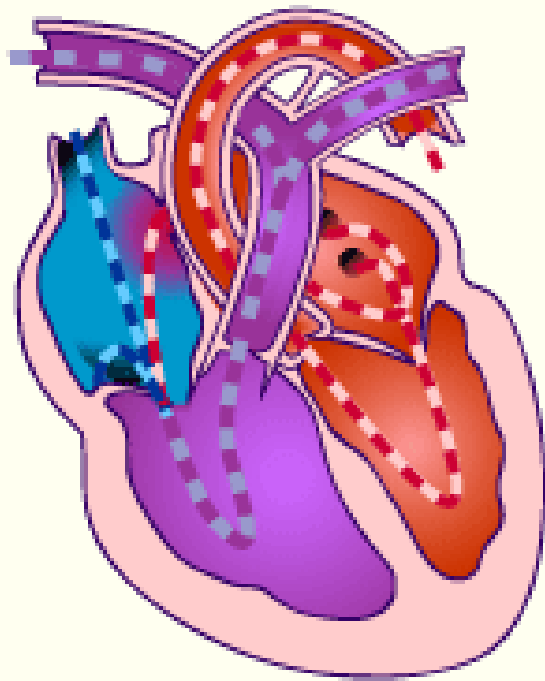
Pediatric cardiologist to read
TeleCardiology consult



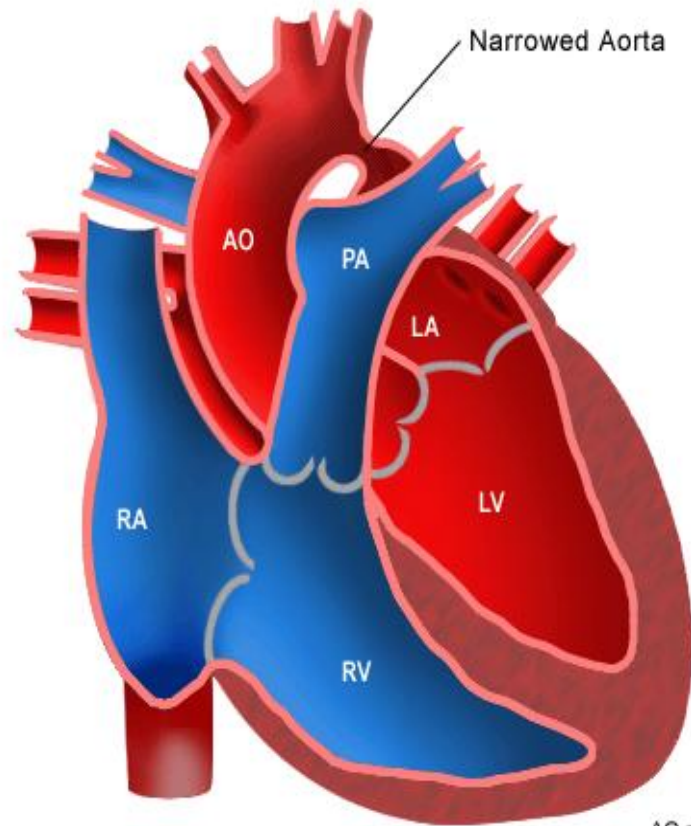
- <http://www.stonybrookphysicians.com/images/Pediatric-Echo-2008.jpg>
- Ucdavis.edu



Will all types of heart defects be picked up with pulse oximetry?

- There is no current screening method that will pick up all heart defects.
- One third of heart defects may not be found on screening (no decrease in oxygen saturations)



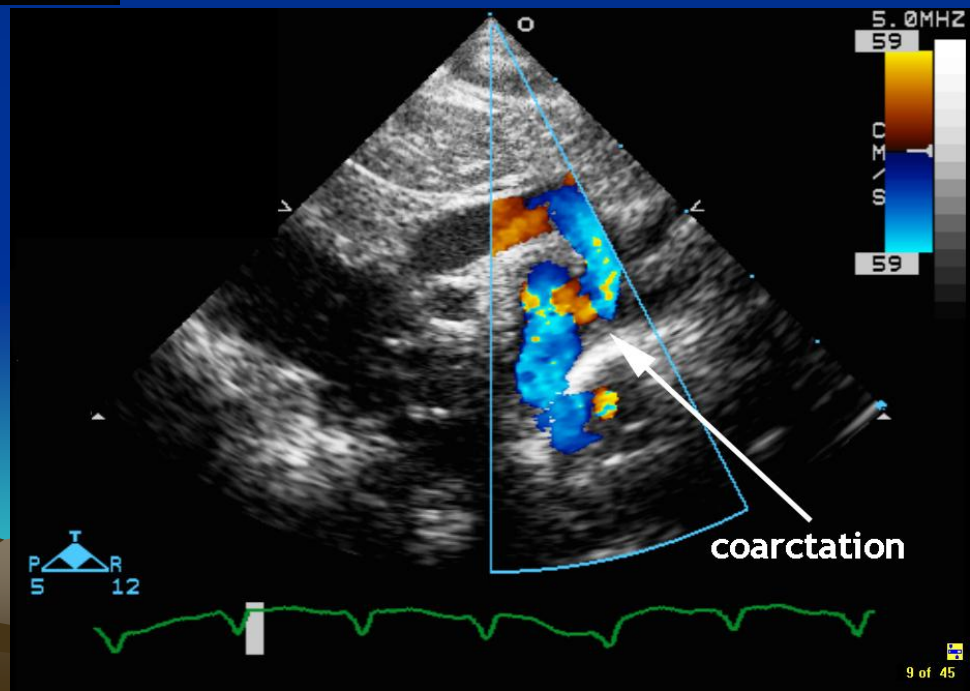
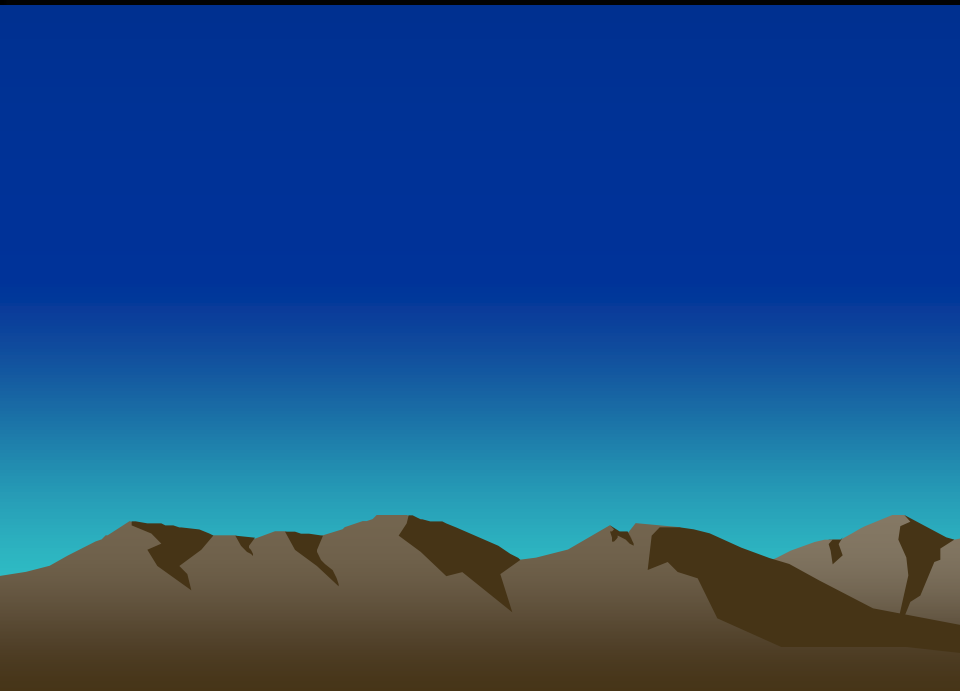
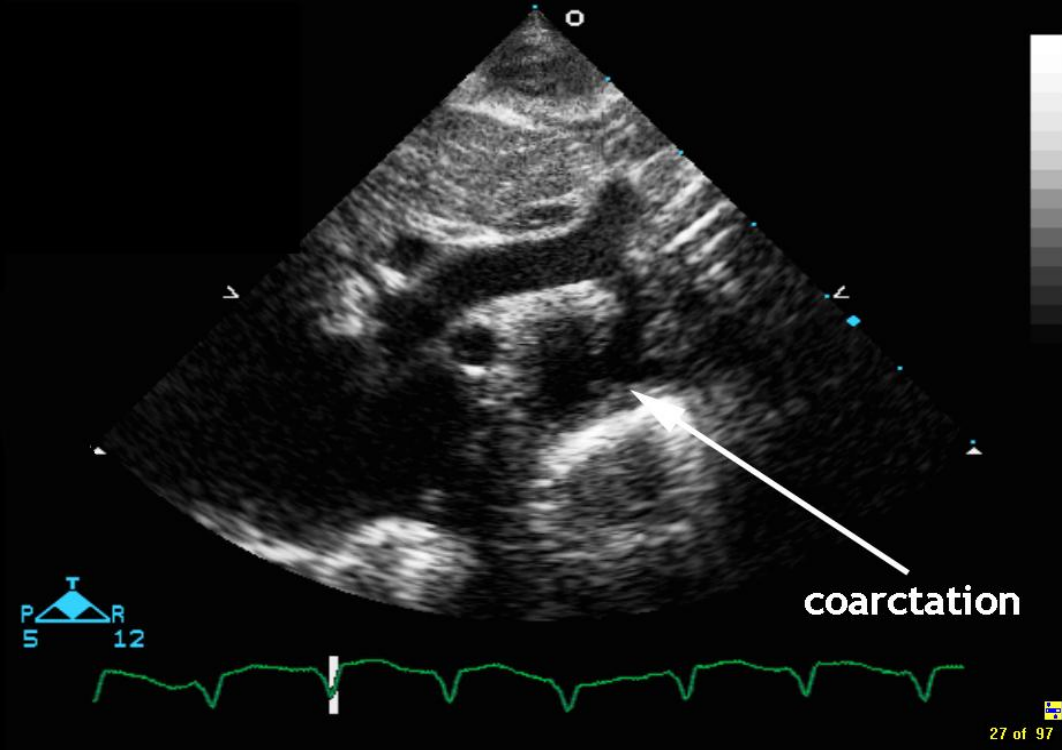
Obstructed L side Coarctation



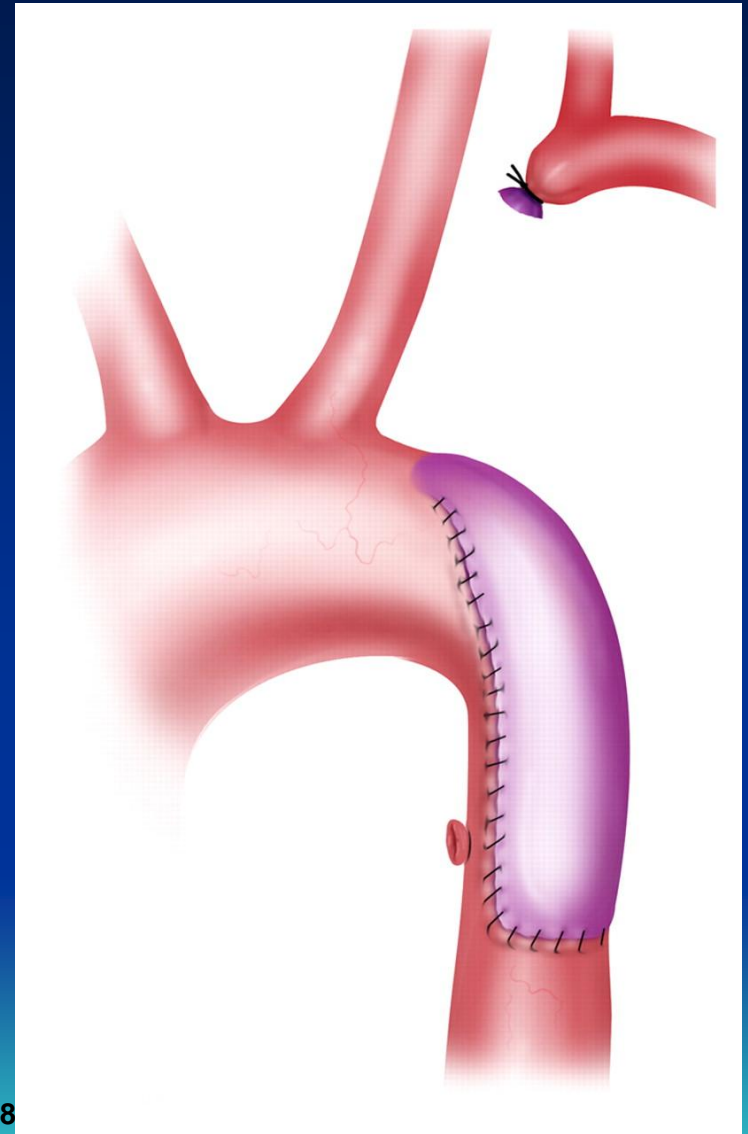
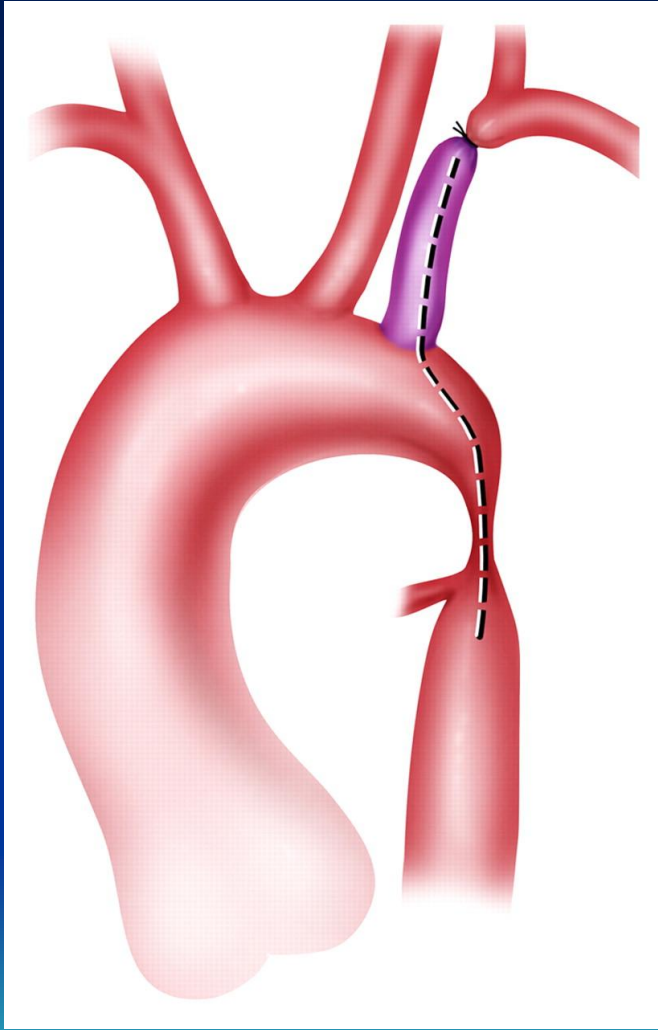
 Oxygen-rich Blood
 Oxygen-poor Blood

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

Doppler Flow



Illustrations show Waldhausen repair of coarctation of aorta.



Gaca A M et al. Radiology 2008;248

Radiology



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References

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- De-Wahl Granelli, A et al (2008). Impact of pulse oximetry screening on the detection of duct dependent congenital heart disease: a Swedish prospective screening study in 39,821 newborns, *BMJ*, 337:a3037.
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- Riede, FT Wörner, C et al (2010). 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*, 169: 975-81.
- www.cdc.gov/ncbddd/pediatricgenetics/pulse
- www.cchdscreeningmap.com
- National Center for Hearing Assessment and Management, Utah State University www.infanthearing.org/legislative/mandates.html

California Legislature AB 1731

Assembly Bill

- (d) The department shall expand statewide screening of newborns as soon as possible to include pulse oximetry screening, when feasible between 24 and 48 hours after birth, for critical congenital heart disease.
- 3/1/12 Sent to Committee on Health
- http://e-lobbyist.com/gaits/view/400634?utm_campaign=rss&guid=7CpLV75Qk7misKkOnLV
[S7A](#) introduced 2/16/12



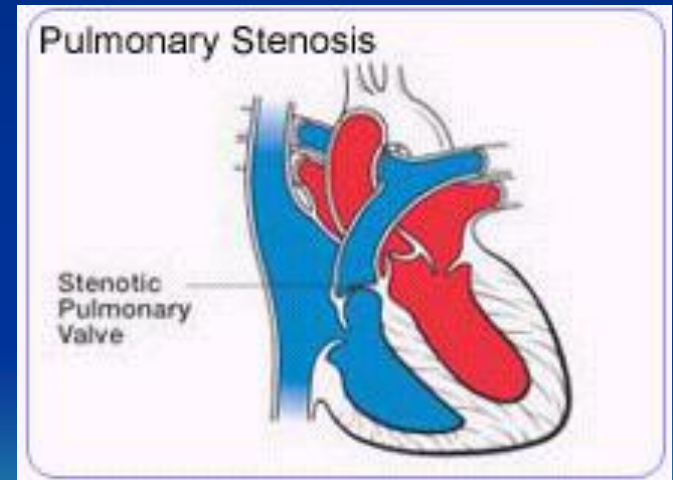
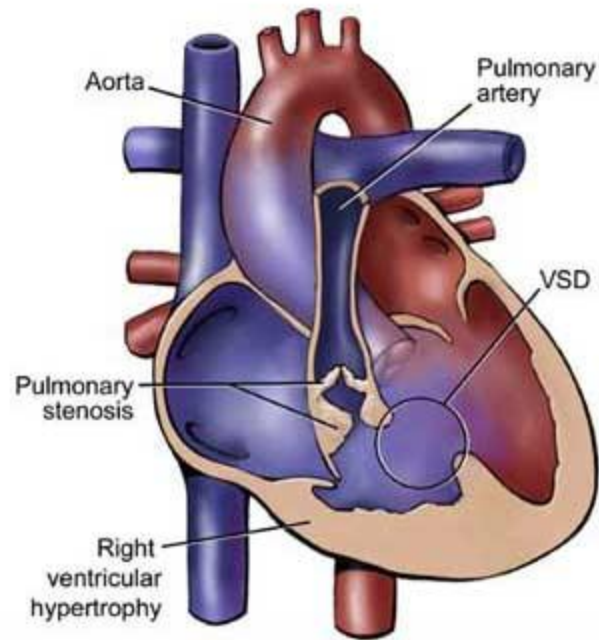




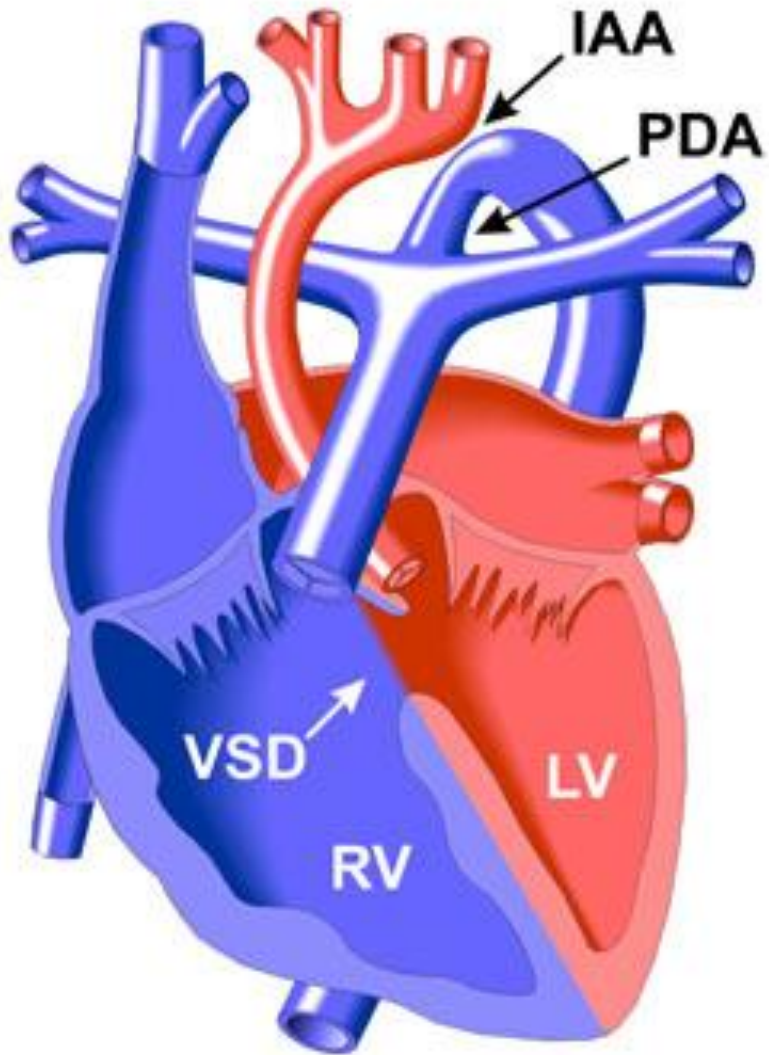
- Lethal (1000 x) heparin dose.
- \$750,000 judgment Cedars-Sinai
- LegalRadar
- 12/18/08
- 11/07 twins

Obstructed R side

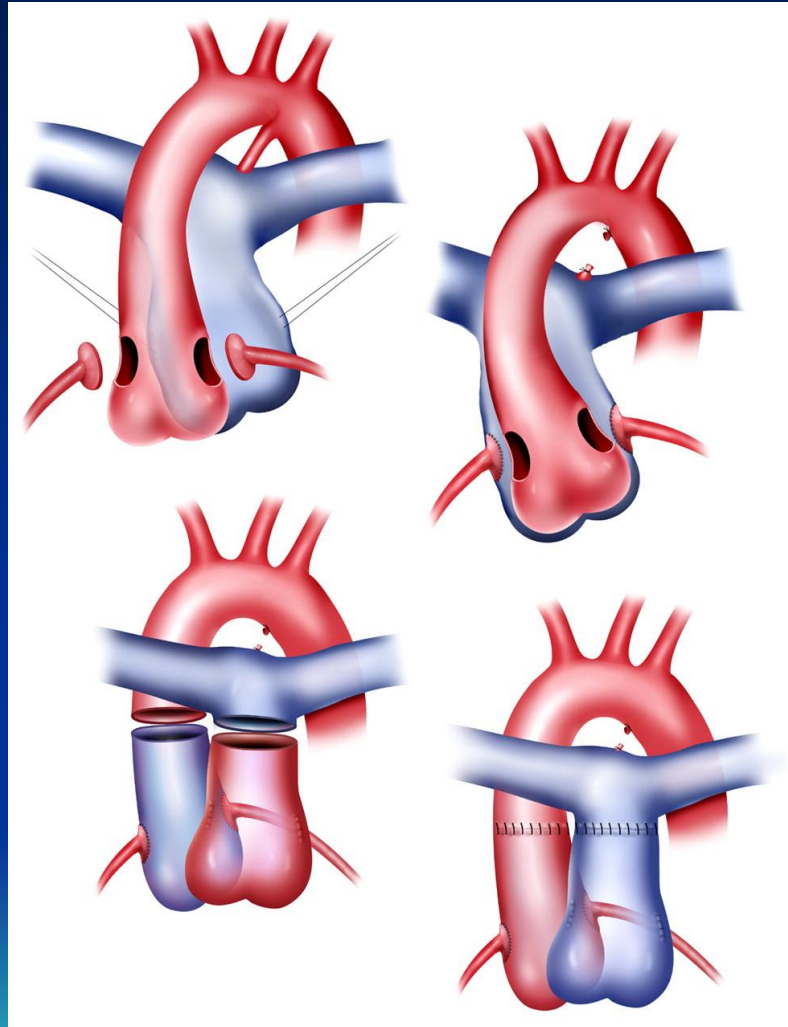
- Pulmonary stenosis (valvular)



Interrupted Aortic Arch



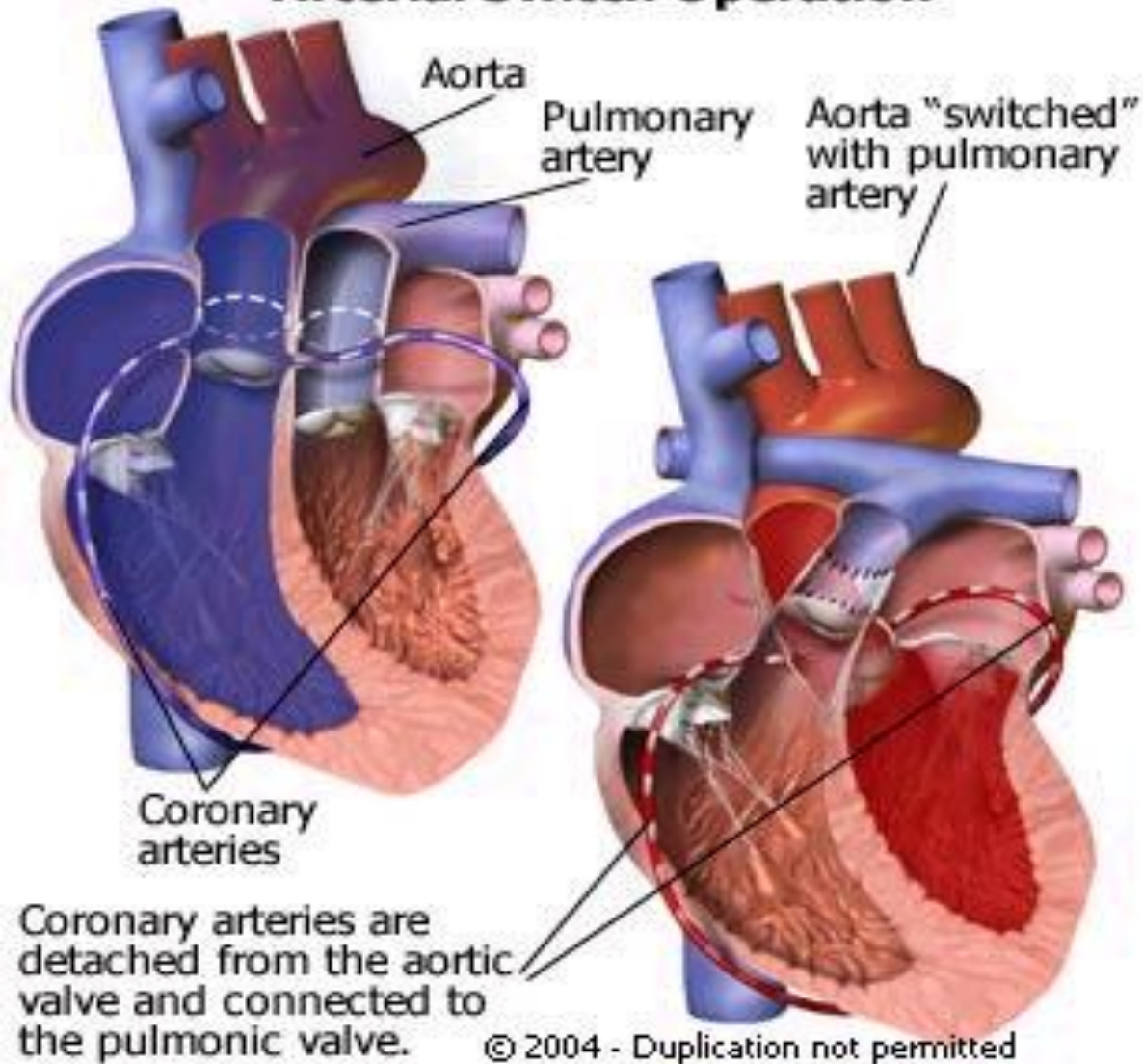
Illustrations of Jatene arterial switch procedure.



Gaca A M et al. Radiology 2008;247:617-631

Radiology

Arterial Switch Operation



Truncus Repair

