OSD
Congenital Heart Disease

Part 2—Cyanotic heart disease, treatment options and post-operative physiology

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I have no conflicts of interest or disclosures to make

Overview/Objectives

- **Act I: Rules of the Game**
  - Explain why some congenital heart conditions lead to congestive heart failure
  - Explain why some congenital heart conditions lead to hypoxemia
  - Explain how some congenital heart conditions can lead to CHF and hypoxemia

- **Act II: Ductal-dependence and the "5 T's"**
  - Describe the concept of ductal-dependence
  - Compare and contrast the most common forms of cyanotic congenital heart disease

- **Act III: Systemic outflow obstruction**
  - Describe at least one type of congenital systemic outflow obstruction
  - Explain common themes between these conditions and the "5-Ts"

- **Act IV: Repair/Treatment options and consequences**
  - List at least 2 general concepts related to the treatment/repair of congenital heart defects
  - Describe the physiologic consequences of single ventricle palliation

ACT 1: Rules of the Game

Let's quickly review the take-home points from Part I of this talk:

- Blood generally follows the path of least resistance
- Remodeling occurs in the face of pressure and volume challenges, which can have adaptive or maladaptive effects
- Pulmonary overcirculation plus left-sided volume overload results in signs and symptoms of congestive heart failure
- Unrepaired left to right shunts can result in Eisenmenger syndrome

ACT 1: Rules of the Game

- Blood generally follows the path of least resistance

- ...and in the case of a ventricular septal defect (VSD) or patent ductus arteriosus (PDA), the path of least resistance is out toward the lungs.

- But what if the resistance is *higher* on the path to the lungs?

Tetralogy of Fallot

- The most common "cyanotic" congenital heart disease (about 1:2,500 live births in U.S.)

Tetralogy of Fallot

- Memorize (for boards):

1. Ventricular septal defect
2. Pulmonary stenosis
3. Overriding aorta
4. Right ventricular hypertrophy

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Tetralogy of Fallot
(what you should really learn)

- ToF is the result of a particular sort of VSD

- The conal septum is anteriorly malaligned, crowding the subpulmonary area

- The aorta is "dragged" anteriorly to sit over the VSD

- RVH results from the pulmonary stenosis and VSD (ventricles respond to pressure challenge with hypertrophy!)

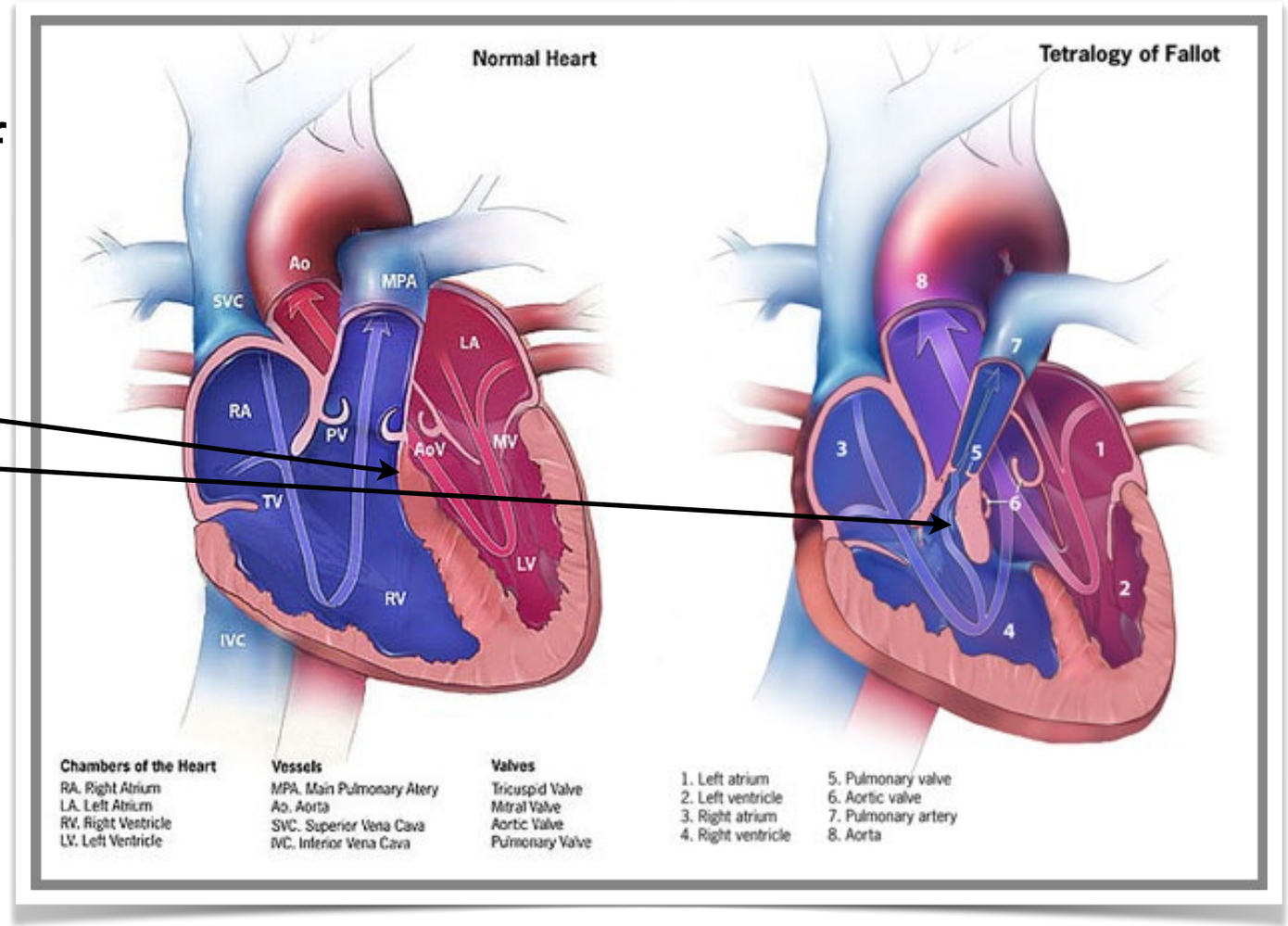

Image from: http://www.heartbirthdefect.com/heart-birth-defects/tetralogy-of-fallot.php

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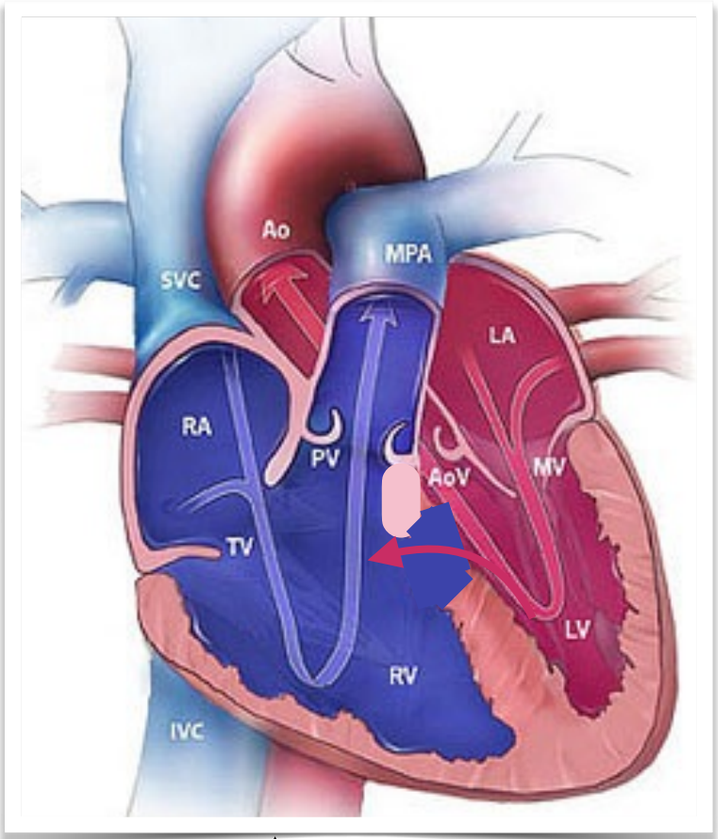
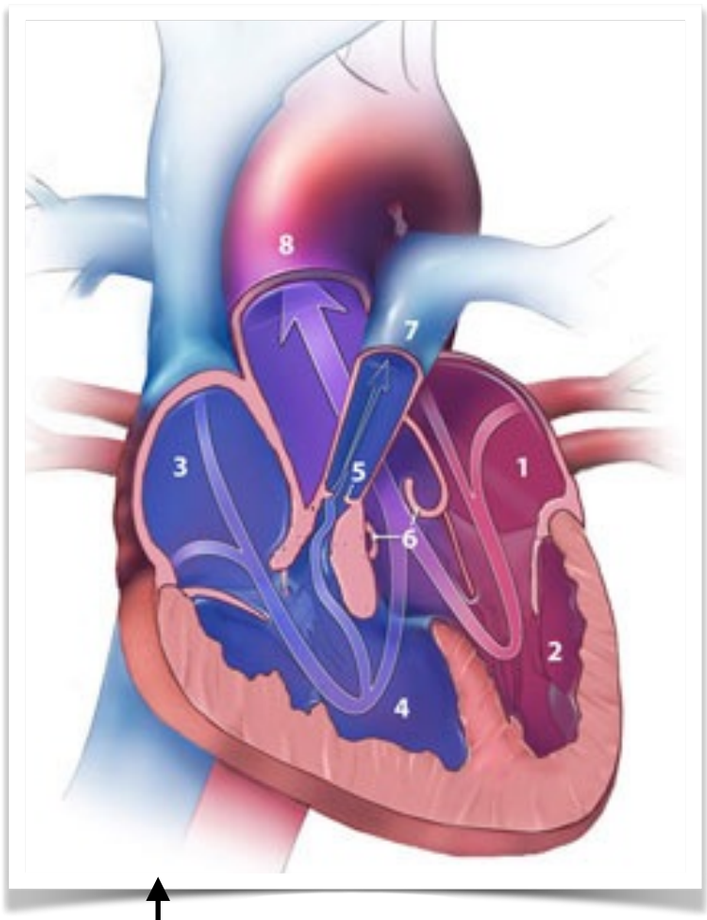
Tetralogy of Fallot exists on a clinical *spectrum* depending on the amount of pulmonary blood flow

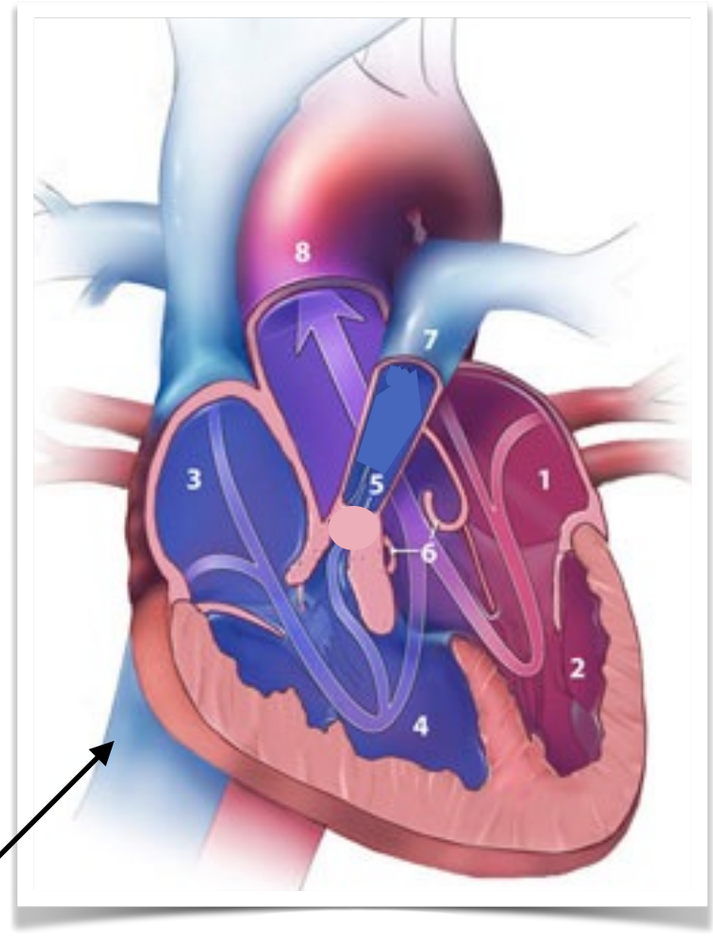


If there is not much pulmonary stenosis, the physiology is that of a simple VSD (pulmonary overcirculation, congestive heart failure)



Often, there is enough pulmonary stenosis to result in right-to-left shunting across the VSD (hypoxemia)

If there is *pulmonary atresia,* how does blood get to the lungs?



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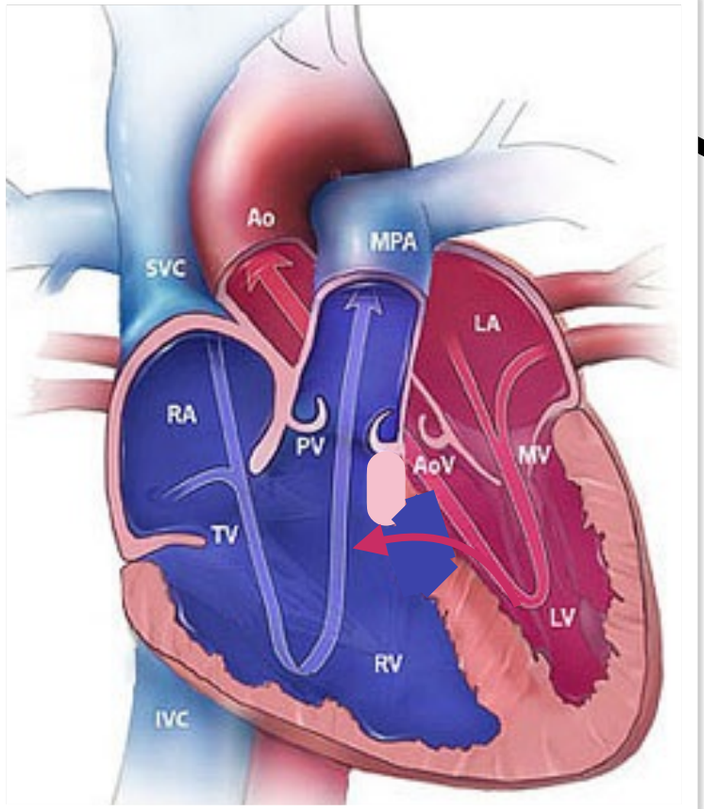
Decreasing Pulmonary Blood Flow



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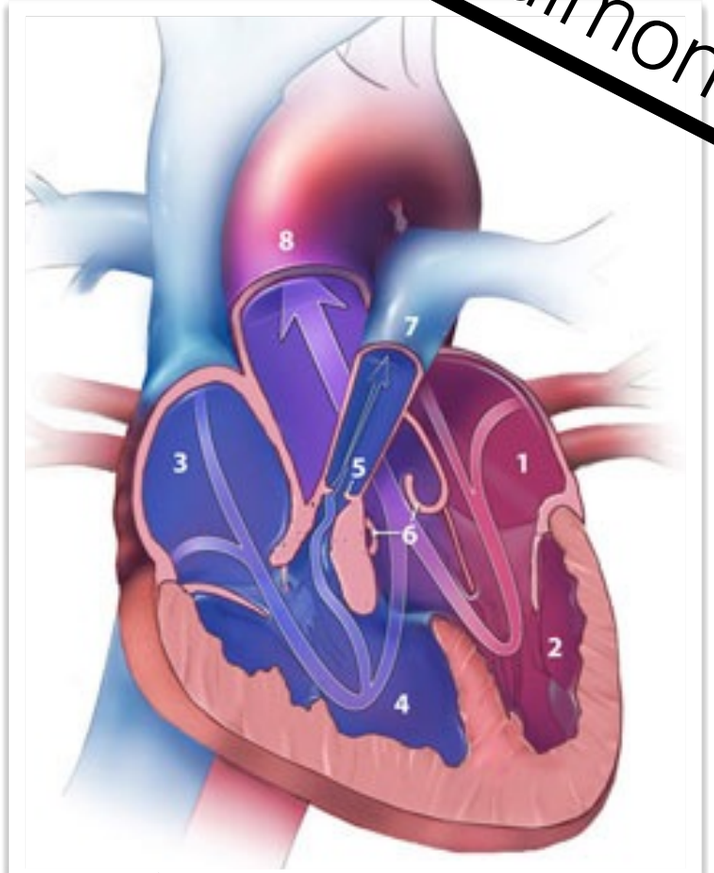

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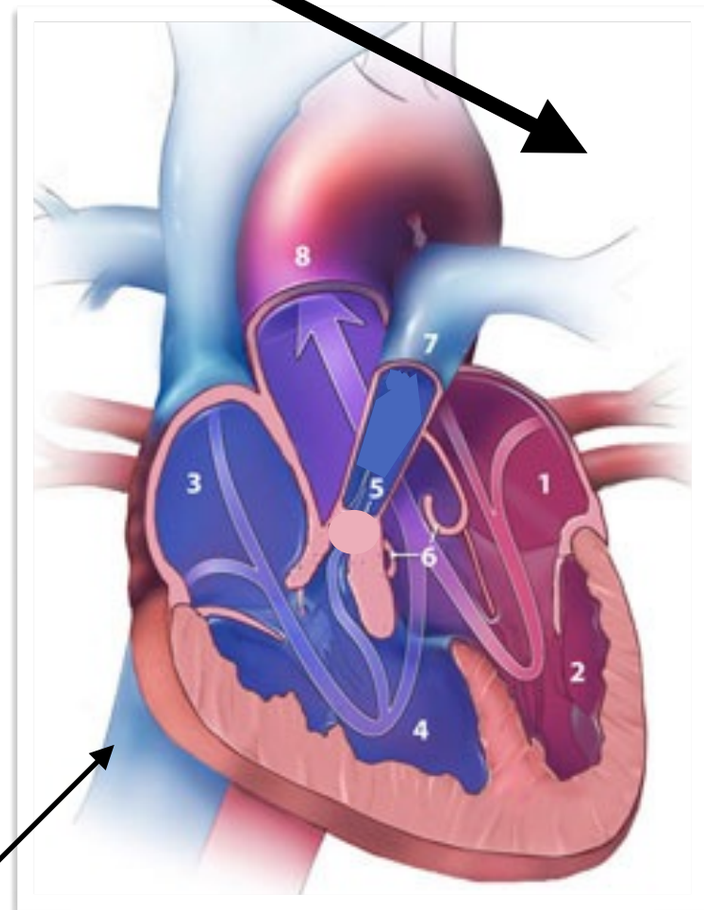

If there is *pulmonary atresia,* how does blood get to the lungs?

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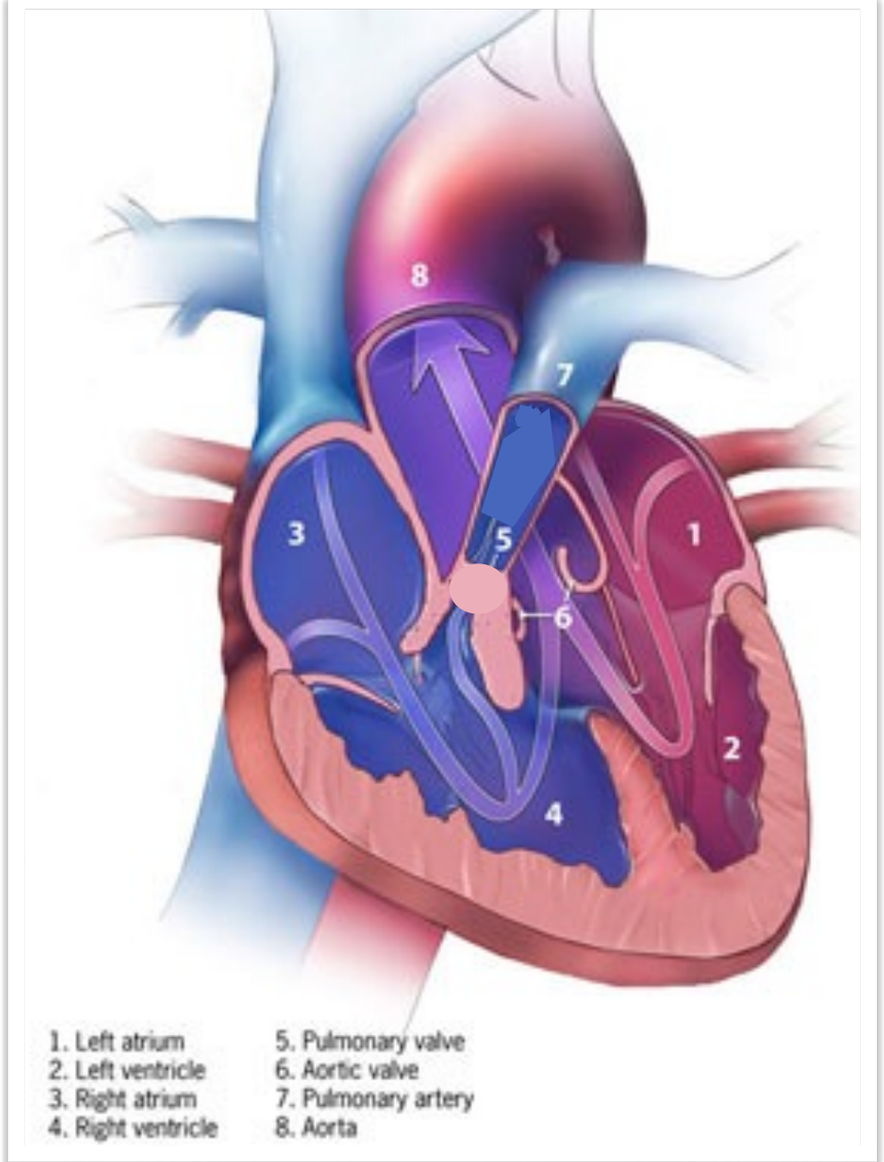
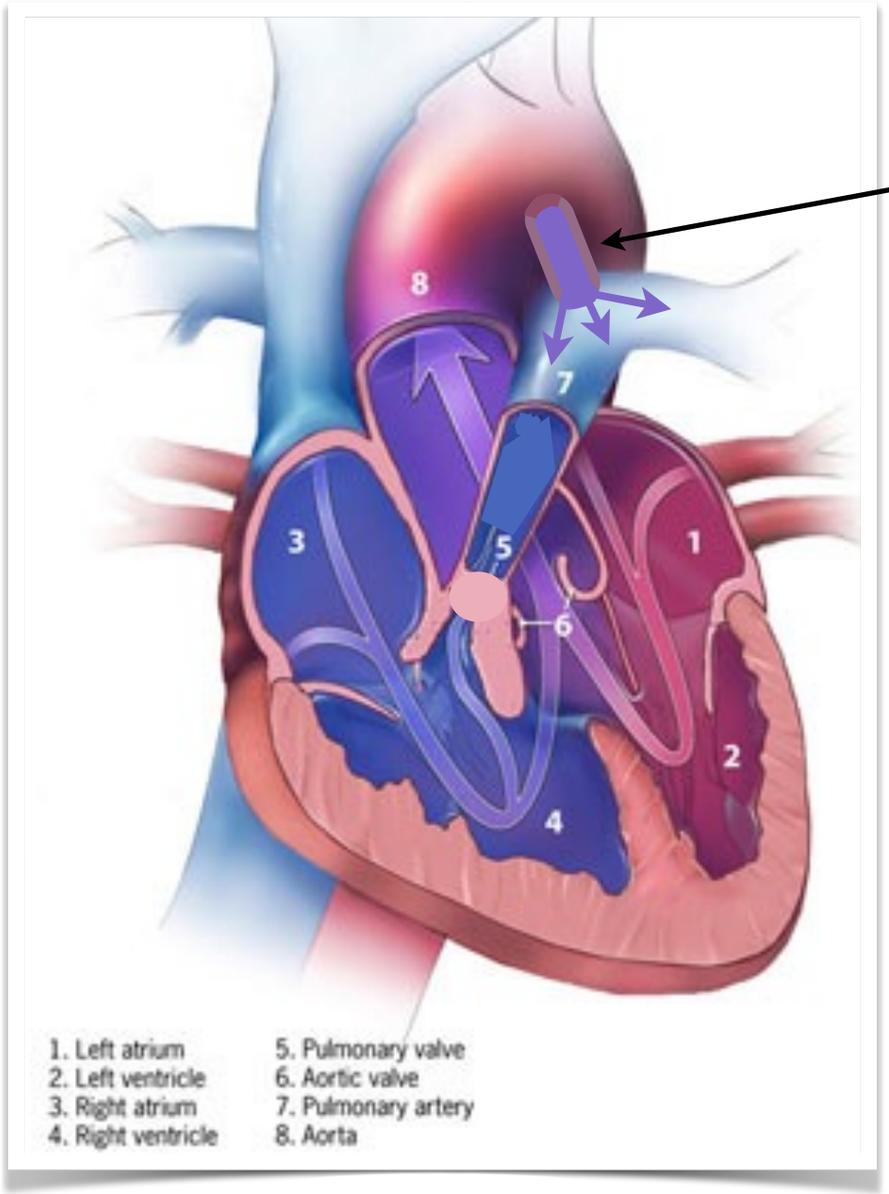
Tetralogy of Fallot with Pulmonary Atresia
--an introduction to "mixing"



- What if there's pulmonary atresia (no valve)?
- There would be ***complete mixing*** of blue and red blood!
- Wait a minute, where is the red blood coming from???
- *If there is pulmonary atresia, how does blood get to the lungs?*

Image from: http://www.heartbirthdefect.com/heart-birth-defects/tetralogy-of-fallot.php

Tetralogy of Fallot with Pulmonary Atresia
--an introduction to "mixing"



- You need a <u>patent ductus arteriosus</u> (PDA)!

- Note a few key points of this physiology:

1) The O2 sat of the Ao and PA are the same
2) The O2 sat of the Ao depends on the quantity of pulmonary blood flow
3) Pulmonary overcirculation can cause CHF
4) No matter how great the pulmonary flow, the patient will still be hypoxemic

<u>WHY?</u>

Image from: http://www.heartbirthdefect.com/heart-birth-defects/tetralogy-of-fallot.php

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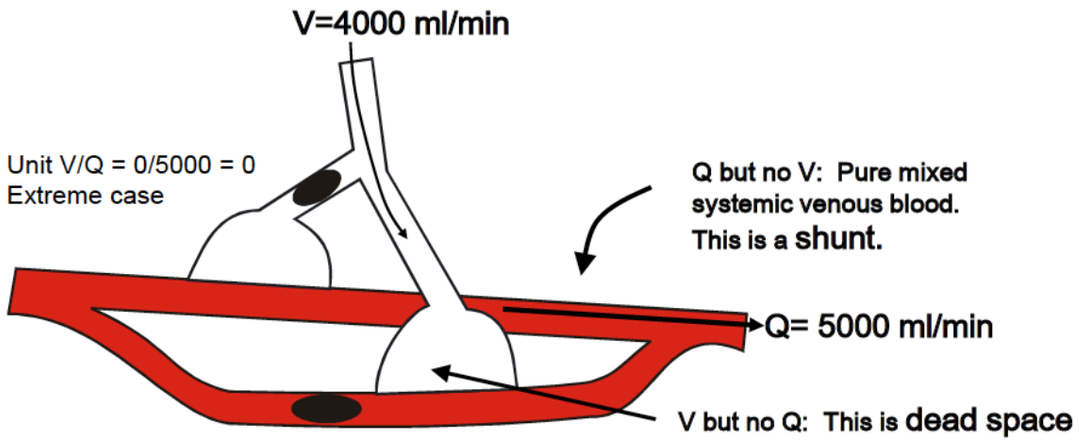
Consider a lung where half of the alveoli units are not perfused
And the other half of the alveoli are not ventilated

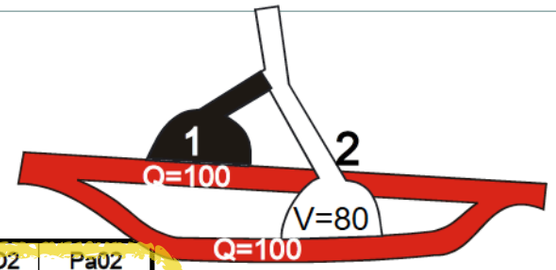


Unit V/Q = 4000/0 = ∞

The lung is both ventilated and perfused... but there is no gas exchange.

For gas exchange to take place, you need appropriate matching of V and Q

Diagnosing a Shunt using 100% O_2



	V_A (ml)	Q (ml)	V/Q	PaCO2	Pa02
Unit 1	0	100	0	45	40
Unit 2	80	100	0.8	40	100

Breathing room air: What's the PaO2 ?

PaO2 = (15 mls + 19.8 mls) / 2= 17.25 vol% => PaO2 = 53 mm Hg

Breathing 100% O_2: Now what's the PaO2 ?

	V_A (ml)	Q (ml)	V/Q	PaCO2	Pa02
Unit 1	0	100	0	45	40
Unit 2	80	100	0.8	40	500

Unit 2 O2 content = (.003 x 500) (=dissolved) + 19.8 (Hb fully sat'd) = 21.6

Average PaO2 = (15 mls + 21.6 mls) / 2 = 18.3 vol% → 65 mm Hg

Slides from: Zayaruzny—V/Q Matching Principles—DSF 2020

Causes of hypoxia

- Hypoxic hypoxia
  - Hypoventilation
  - Diffusion abnormalities
  - Shunt
  - V/Q mismatching
- Anemic hypoxia
- Hypoperfusion hypoxia
- Histotoxic hypoxia

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Congenital Heart Disease Rule #4:

In R to L shunts and "mixing" lesions…

- Hypoxemia is *always* present to some degree
- The hypoxemia cannot be overcome with supplemental oxygen

Act 1:Review

- In congenital heart problems, blood tends to follow the path of least resistance
  - If the path of least resistance is out to the lungs, *pulmonary overcirculation and CHF may result* (CHD Rule #3)
  - If there is a shunt and the path of least resistance is to the systemic circulation, there will be mixed blue and red blood to the body and *hypoxemia occurs* (CHD Rule #4)
    - this hypoxemia cannot be overcome by administering extra O2

Act 1:Review continued

- Congenital heart diseases often exist on a *clinical spectrum* depending on how much pulmonary blood flow there is

- If blood can't get directly out to the lungs through the pulmonary valve/ pulmonary artery, a PDA is necessary to supply the lungs with blood

Act II: Ductal-dependence and the 5 T's

- Let's pick up where we left off with tetralogy of Fallot—the concept of ductal-dependence

- Then we'll look at more examples of mixing

Ductal-dependence:

An important concept in congenital heart disease

- If there is ever a situation where blood cannot flow in sufficient quantity to the systemic or pulmonary vasculature, a PDA is necessary to allow supplemental flow to that bed

- In the newborn, we can maintain patency of the PDA with the use of *prostaglandin E1*

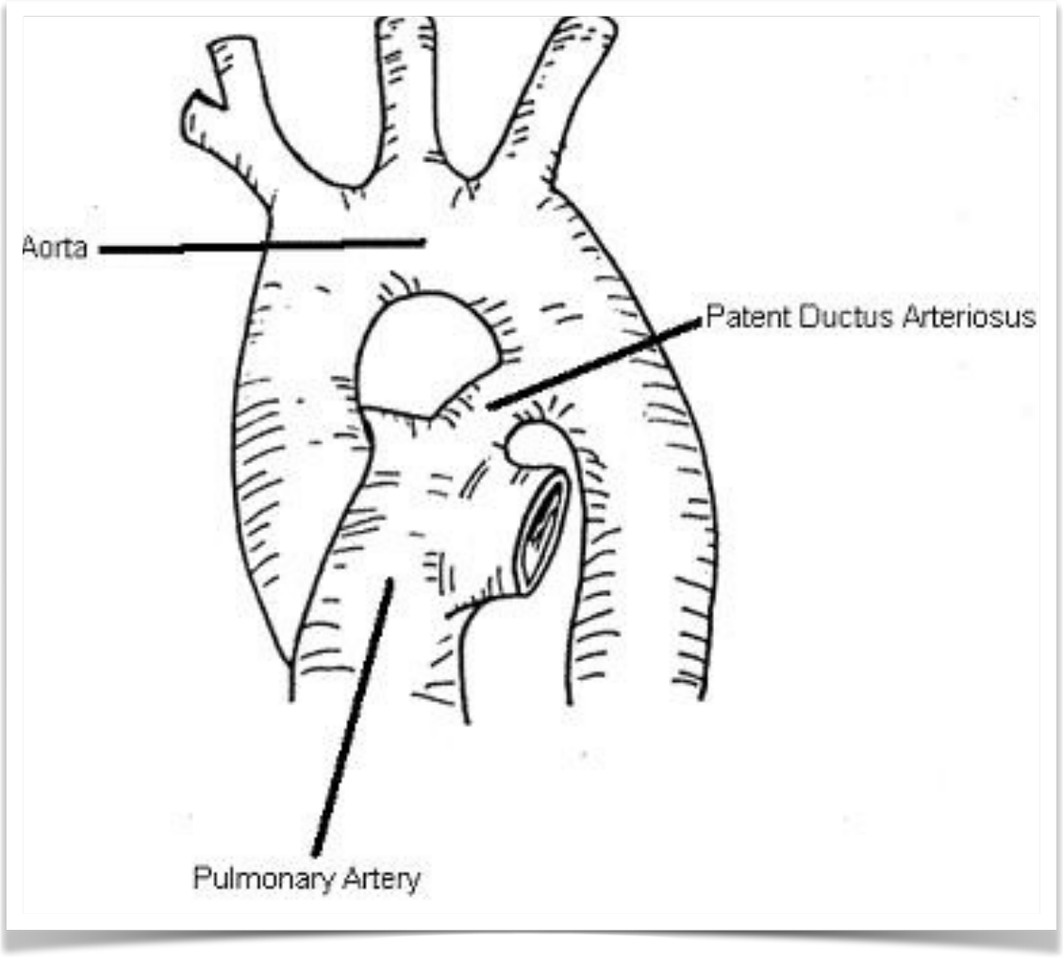

Image from: http://easypediatrics.com/wp-content/uploads/2012/07/Patent_Ductus_Arteriosus.jpg

The 5 "T's" of cyanotic CHD

Tetralogy of Fallot

Tricuspid Atresia

Truncus Arteriosus

Transposition of the Great Arteries

Total Anomalous Pulmonary Venous Return

Memorize these for purposes of the Boards

This is a very incomplete list, however!

It is much more important to grasp the general concepts that dictate the pathophysiology of these defects

Tricuspid Atresia

--a natural progression (conceptually) from ToF

- Tricuspid valve fails to form
- Blue blood must flow R-->L across a PFO/ ASD
- Therefore *complete mixing* of blue and red blood
- Some patients have a VSD, allowing blood to go from LV ---> RV ---> lungs
- If there is no VSD or there is pulmonary atresia, how does blood to get to the lungs?

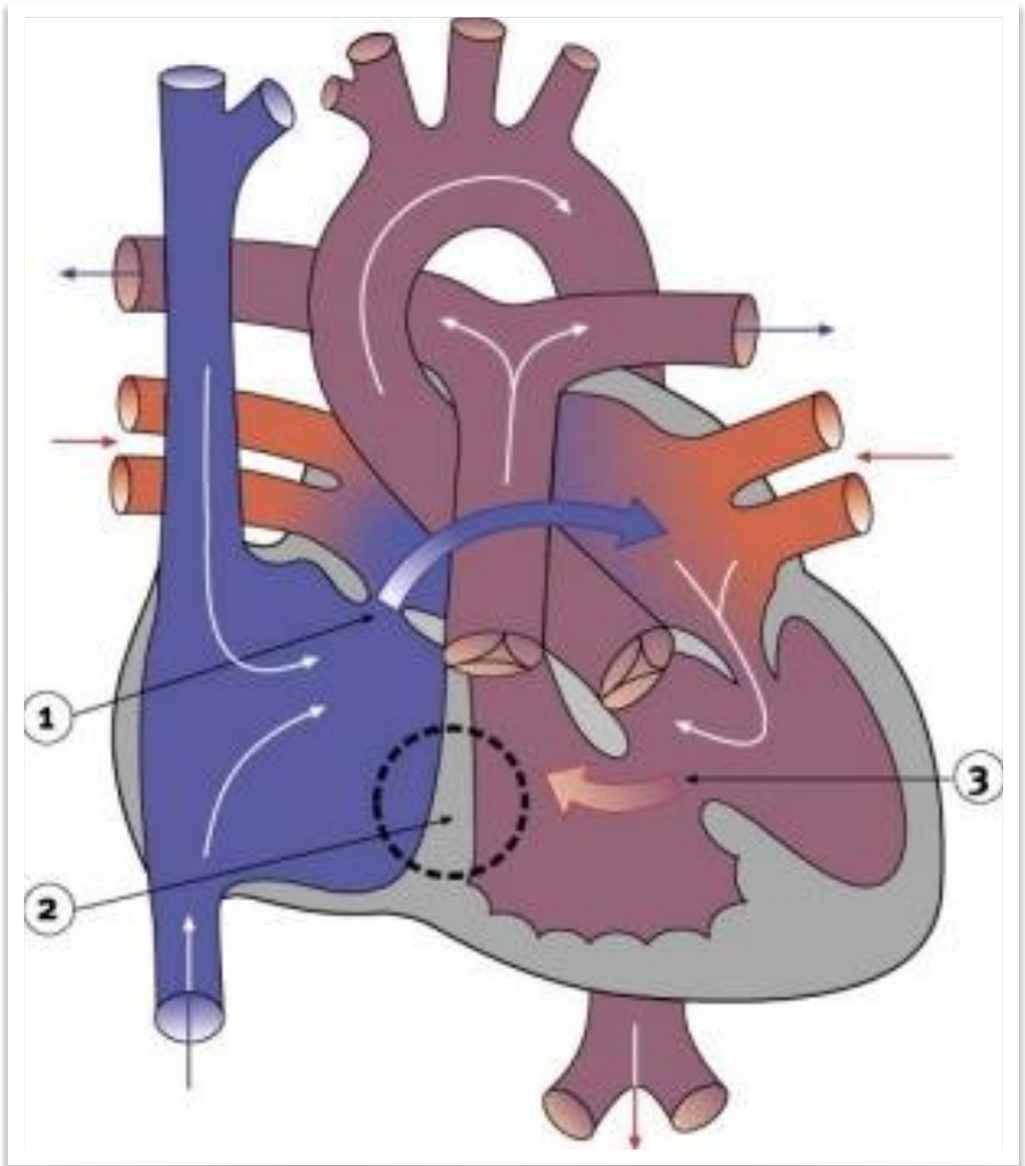

Image from: http://www.pediatriccardiacinquest.mb.ca/ch02/tricuspid.html

Tricuspid Atresia

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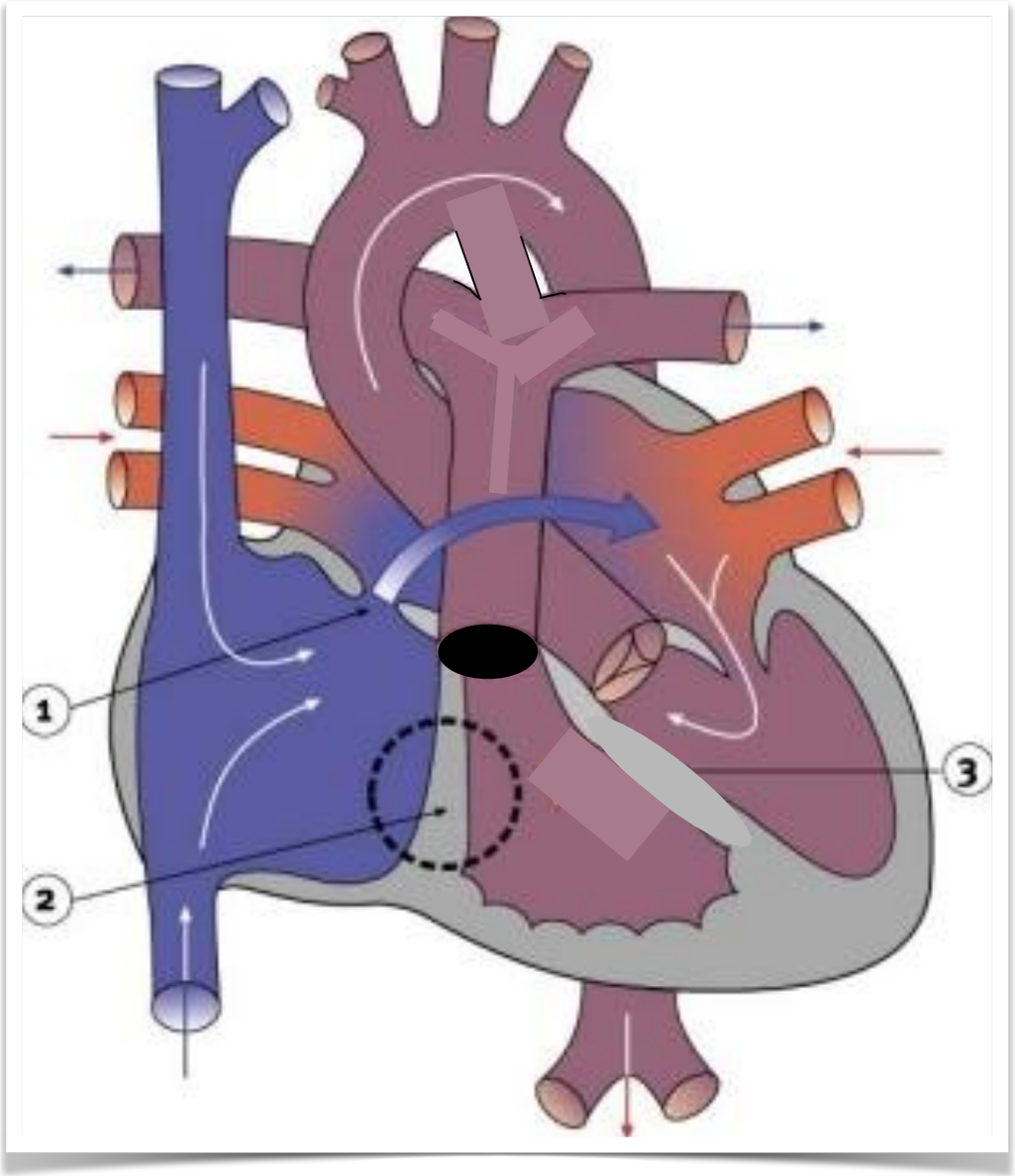


Image from: http://www.pediatriccardiacinquest.mb.ca/ch02/tricuspid.html

- What if there is no VSD or if there is pulmonary atresia?
- You need a patent ductus arteriosus (PDA)!
- Note a few key points of this physiology:

1) The O2 sat of the Ao and PA are the same
2) The O2 sat of the Ao depends on the quantity of pulmonary blood flow
3) Pulmonary overcirculation can cause CHF
4) No matter how great the pulmonary flow, the patient will still be hypoxemic

Does this sound familiar?

Truncus Arteriosus

--a failure of septation

- Failure of septation of outflow tracts and great arteries
- Therefore *complete mixing* of blue and red blood
- Note a few key points of this physiology:

1) The O2 sat of the Ao and PA are the same
2) The O2 sat of the Ao depends on the quantity of pulmonary blood flow
3) Pulmonary overcirculation can cause CHF
4) No matter how great the pulmonary flow, the patient will still be hypoxemic

- *Since blood follows the path of least resistance, what symptoms do you think this patient will have?*

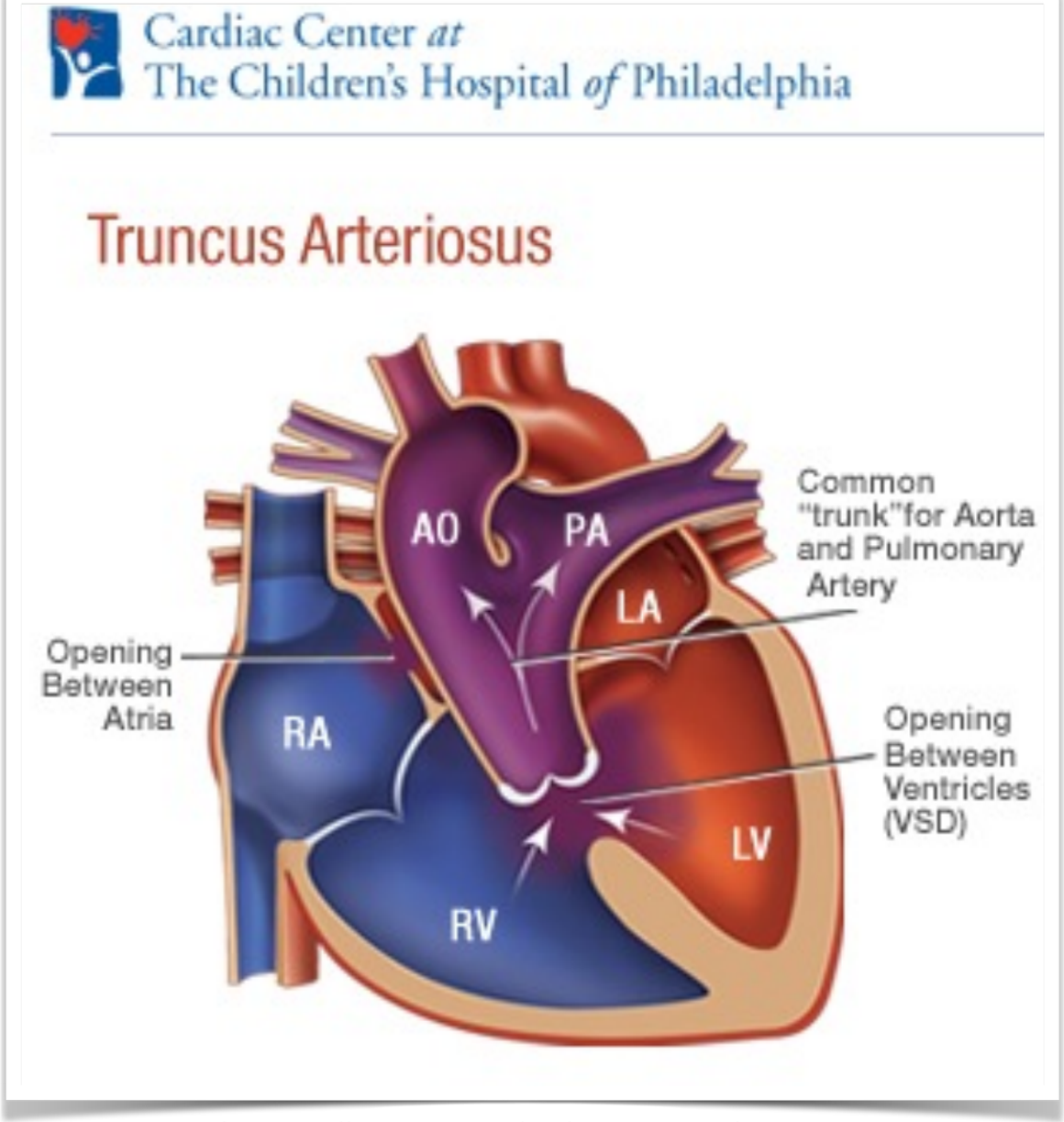


Image from: http://www.chop.edu/img/cardiac-center/truncus-arteriosus.html

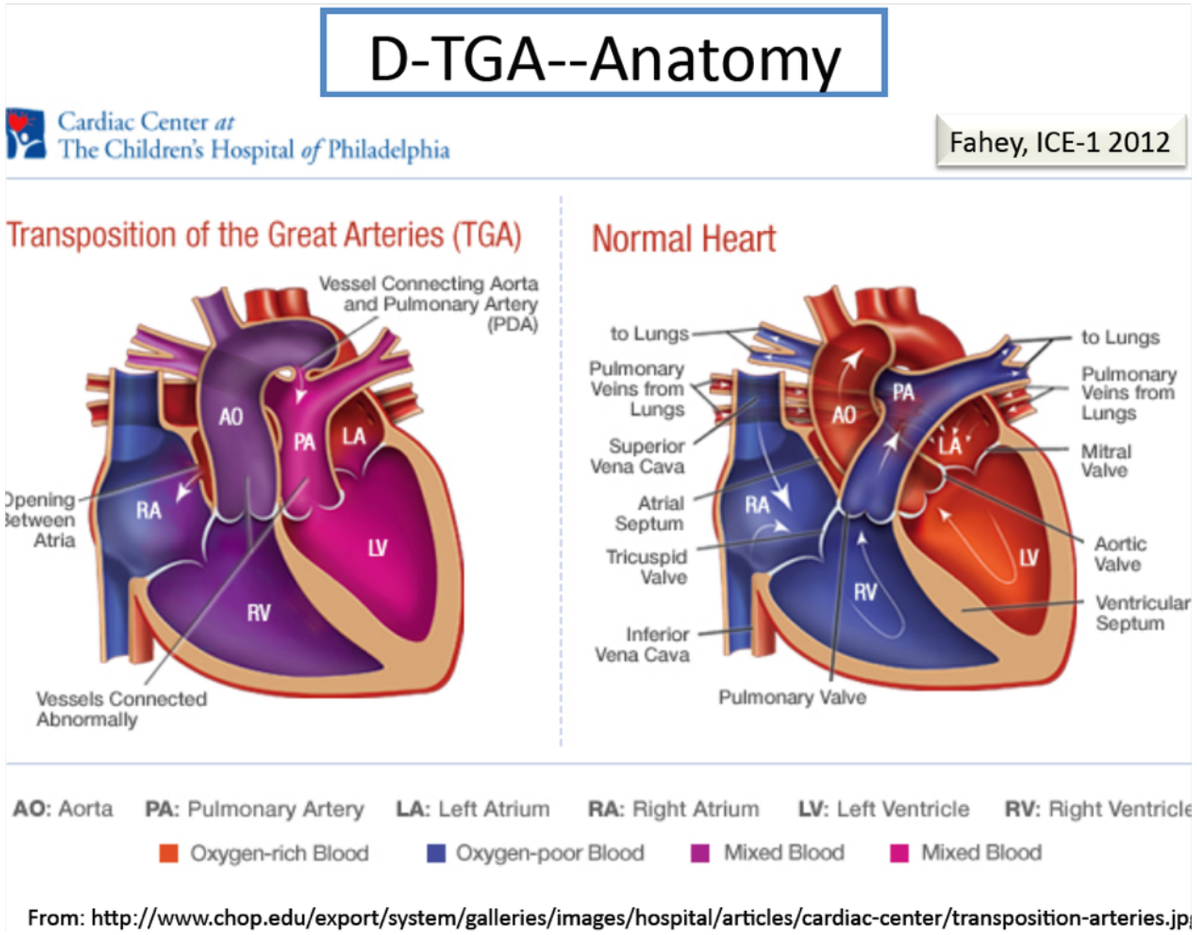
Transposition of the Great Arteries (TGA)

(You've learned about this before!)

D-TGA--Anatomy

Cardiac Center *at* The Children's Hospital *of* Philadelphia

Fahey, ICE-1 2012

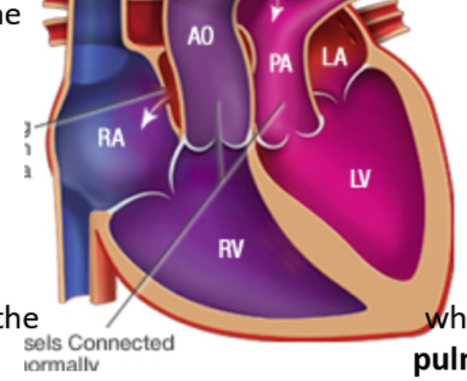


From: http://www.chop.edu/export/system/galleries/images/hospital/articles/cardiac-center/transposition-arteries.jp

Anatomy and physiology of D-TGA...

Deoxygenated (blue) blood returns from the body to the right atrium

...goes to the **right ventricle** where it is pumped through the **aorta** to the body...



The blood is oxygenated and returns (now red) to the left atrium...

...and goes to the **left ventricle** where it is pumped out the **pulmonary artery** and to the lungs...

Fahey, ICE-1 2012

Circulations "in parallel"

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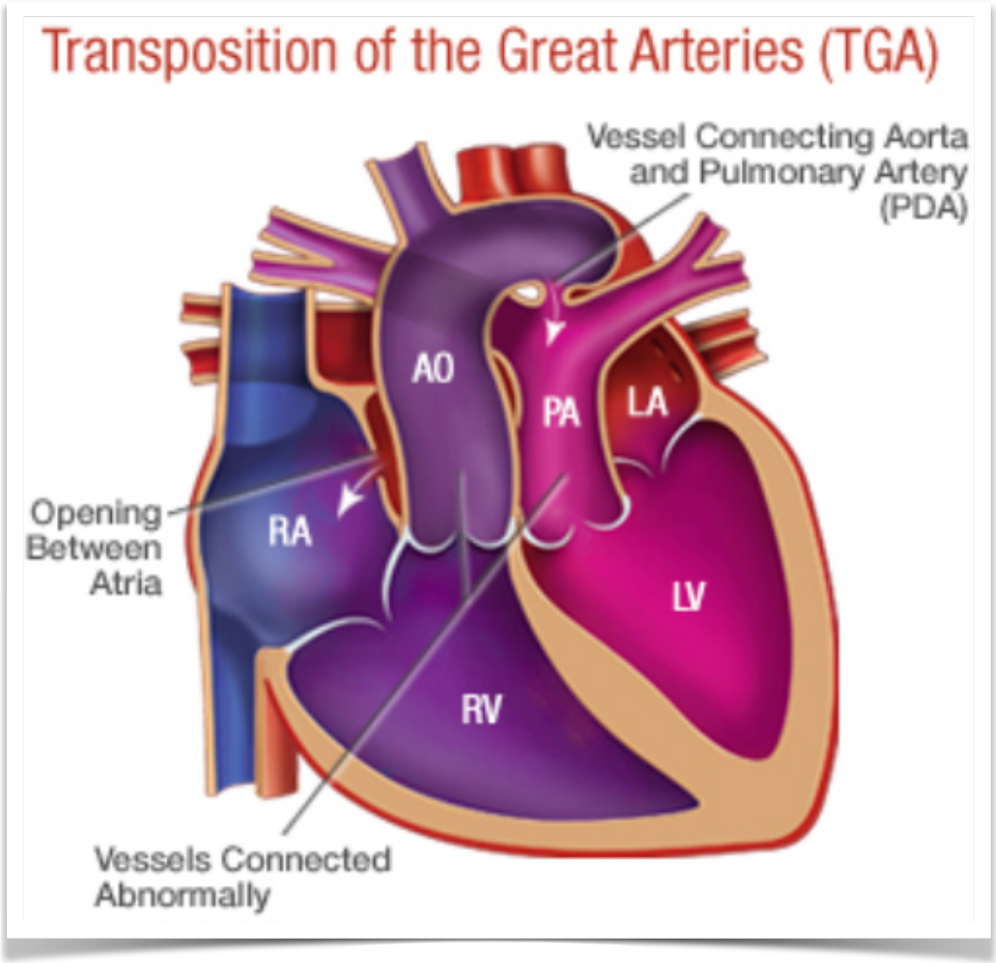
Transposition of the Great Arteries (TGA)

(You've learned about this before!)

- TGA poses a very specific physiologic problem; *circulations in parallel*
- This is a problem unique to TGA
- The short-term solution is to get the circulations to mix more!
- There are 3 places this can occur:
  1) Atrial septal defect
  2) Ventricular septal defect
  3) Patent ductus arteriosus



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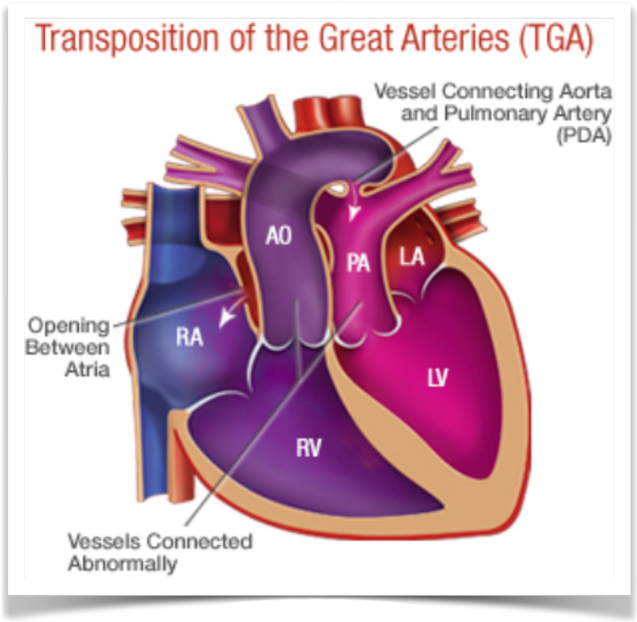
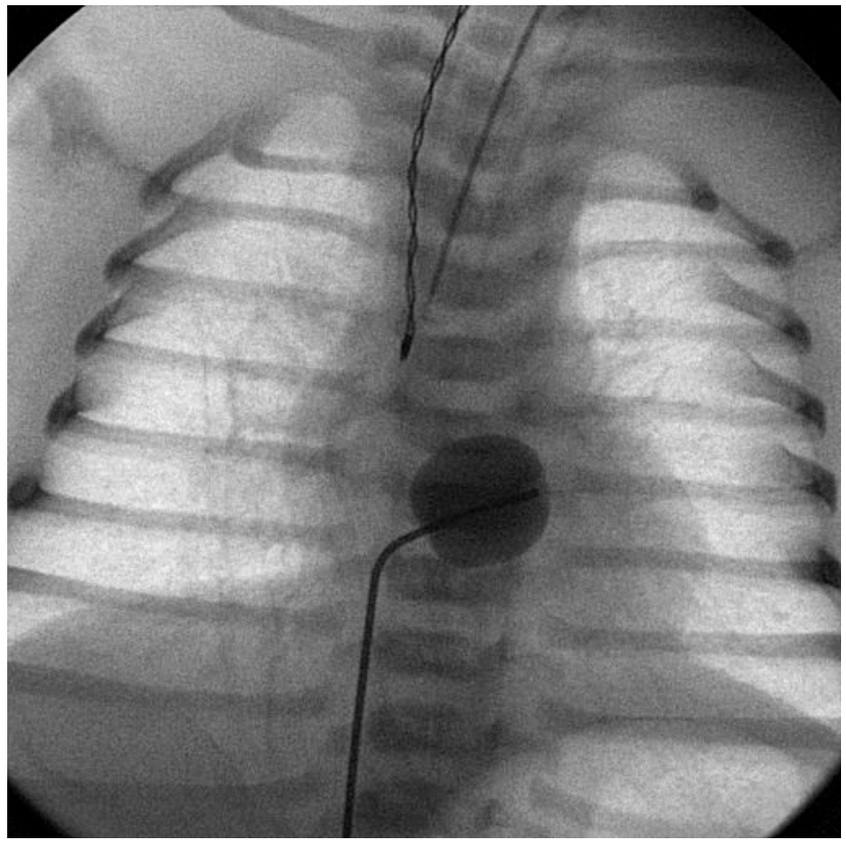
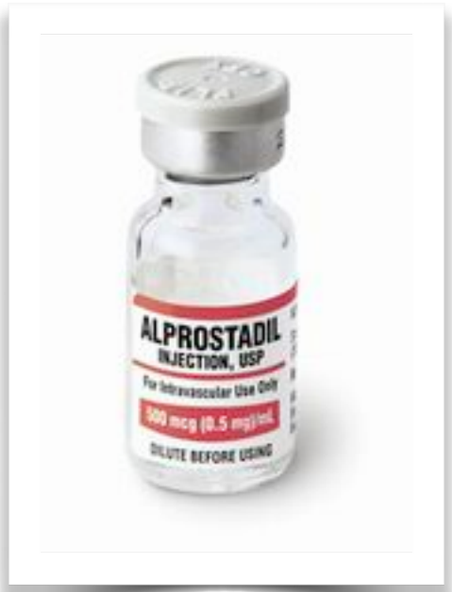
Transposition of the Great Arteries (TGA)

(You've learned about this before!)



- What if there is inadequate mixing?
- You'd better do something about it!





Total Anomalous Pulmonary Venous Return

(TAPVR)

- Pulmonary veins drain to a *confluence* that does not attach to the left atrium

- The confluence drains to a systemic (blue) vein via a *"vertical vein"*
  - Supracardiac (e.g. innominate vein)
  - Cardiac (e.g. coronary sinus)
  - Infracardiac (e.g. inferior vena cava)

- Therefore *complete mixing* of blue and red blood

- If there is no red blood coming back to the left atrium, what goes out the aorta?
  - what does this patient need to survive (one of two things)?

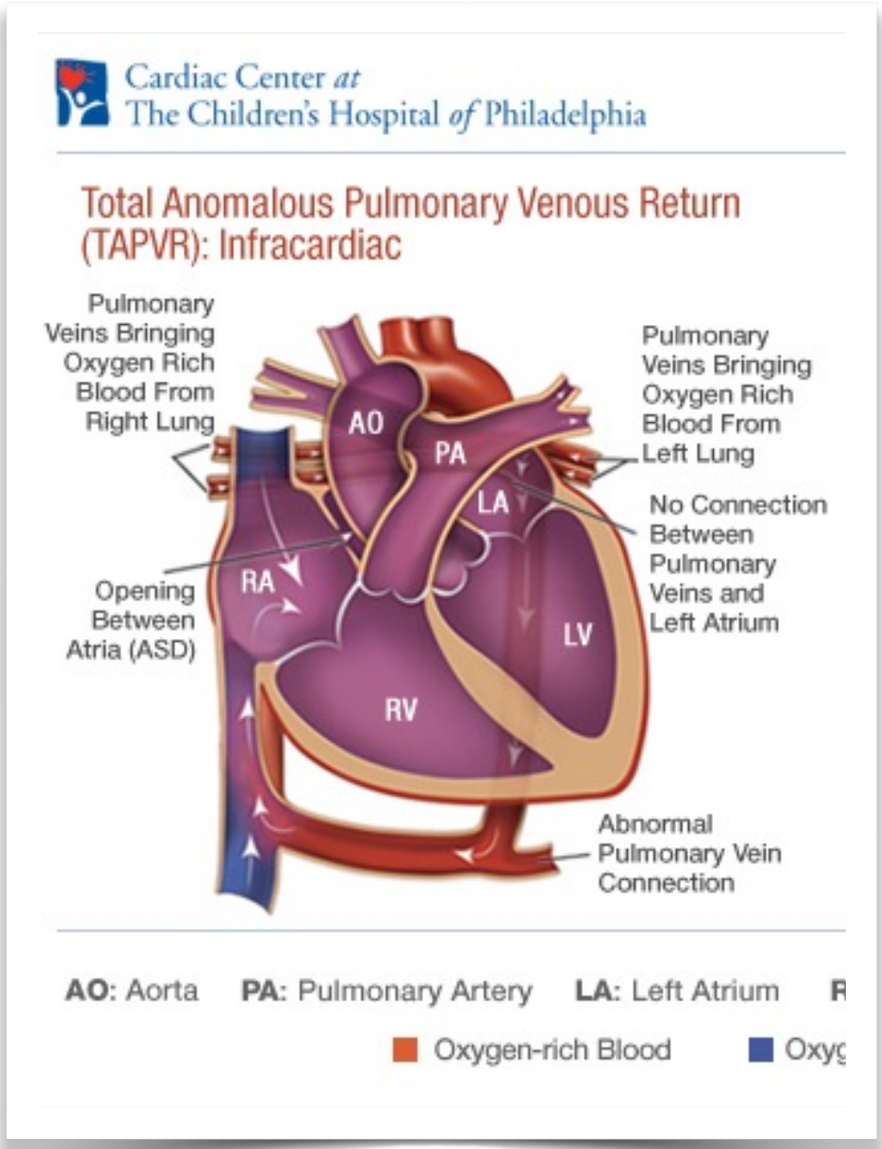


Image from: http://www.chop.edu/img/cardiac-center/tapvr-infracardiac.html

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Total Anomalous Pulmonary Venous Return

(TAPVR)

Blood needs to get to the left side of the heart (via ASD), or out to the Ao (via PDA)

Complete mixing-->hypoxemia

(Maybe) Pulmonary venous obstruction-->CHF

Fixing this requires attaching the confluence to the left atrium, and ligating the vertical vein!

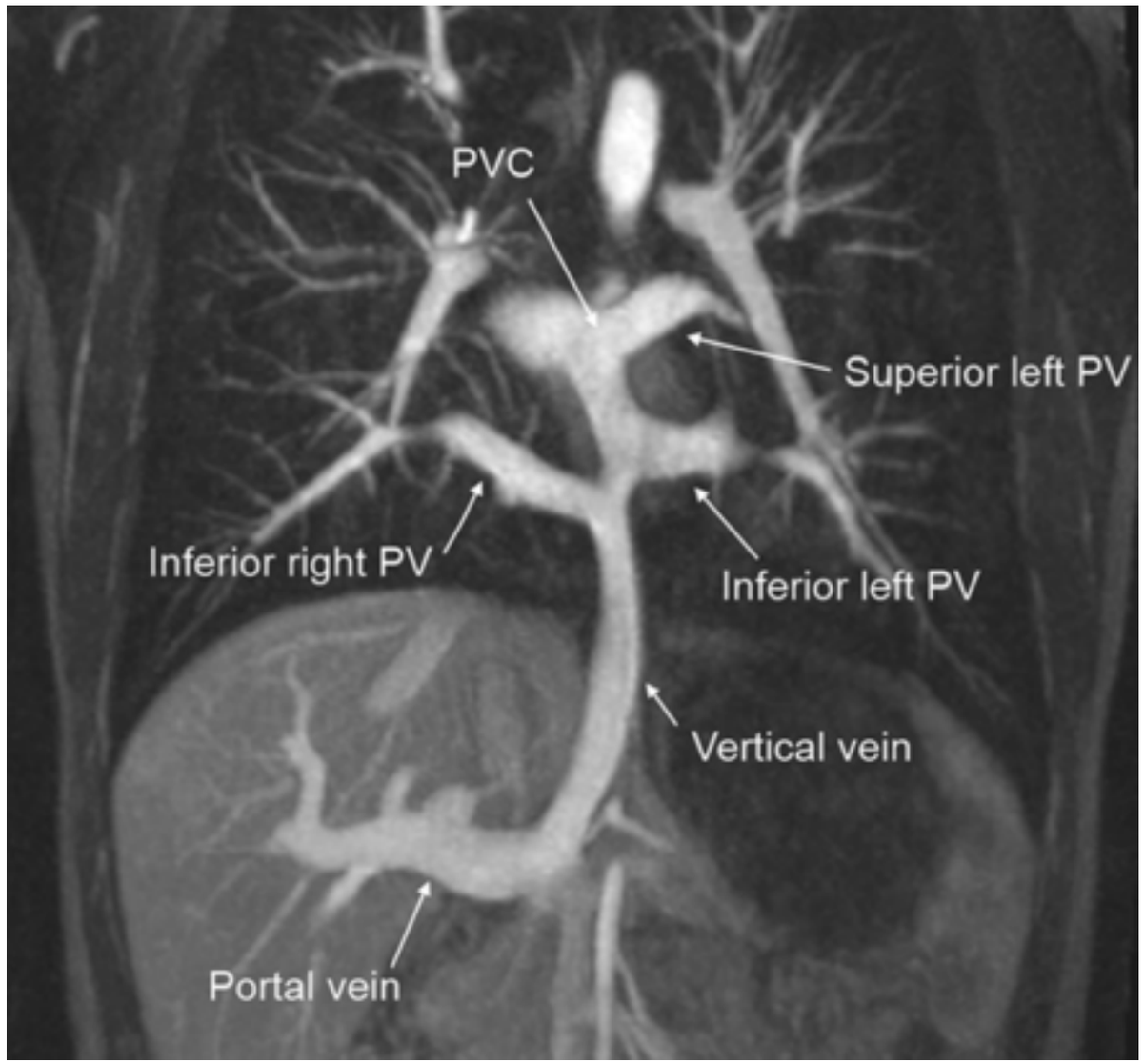
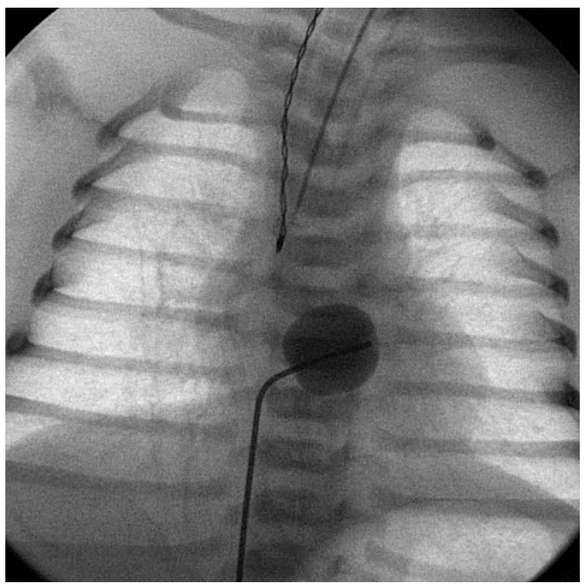


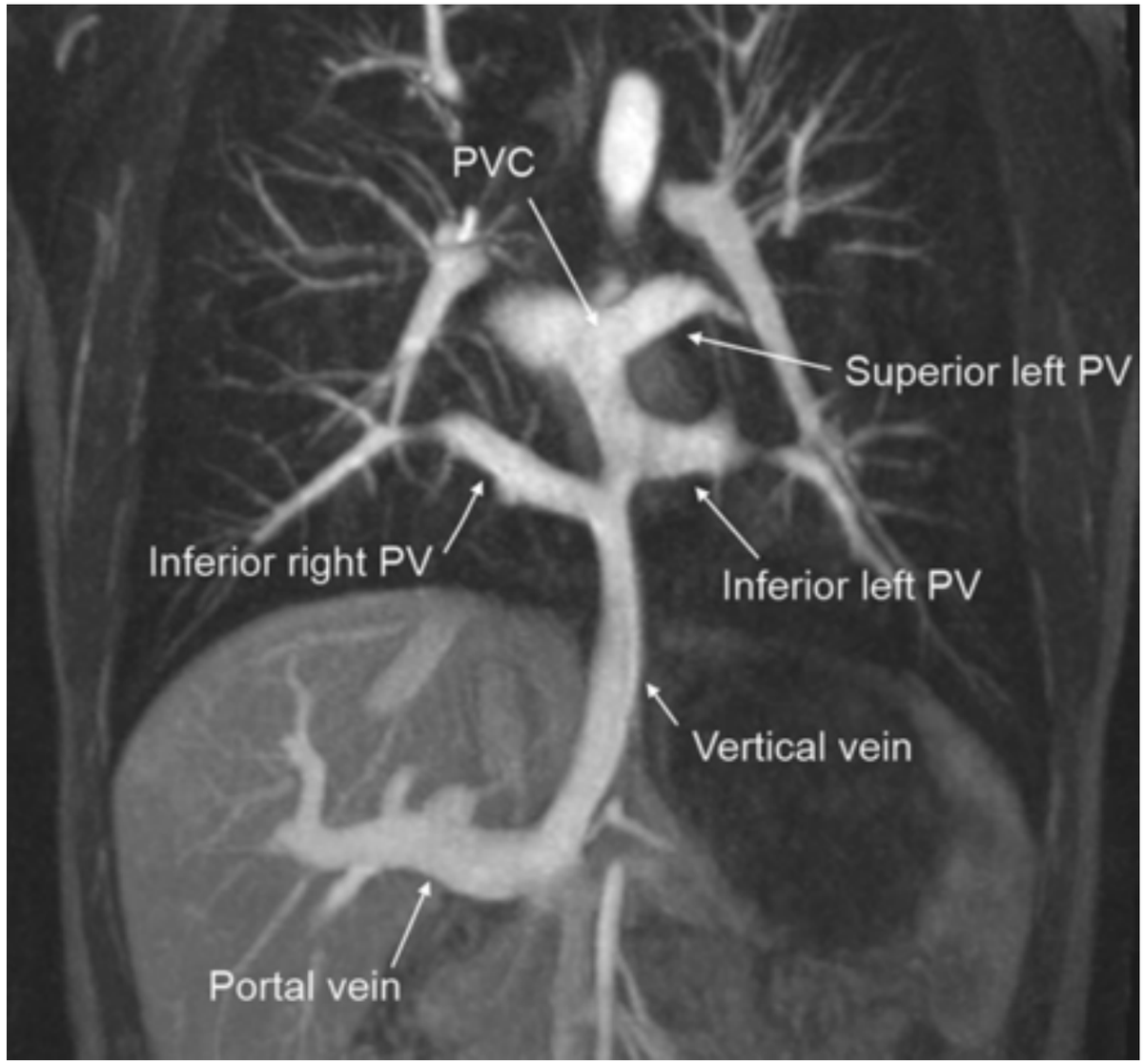
Image from: Circulation. January 15, 2013 vol. 127 no. 2 258-259

Total Anomalous Pulmonary Venous Return

(TAPVR)

Prior to surgical repair, however, is there anything we can do to stabilize the patient?





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Act II Review: 5T's and ductal dependence

- Cyanotic congenital heart diseases often exist on a *clinical spectrum*, much like we saw in the case of tetralogy of Fallot

- Many cyanotic congenital heart diseases share common themes like *ductal-dependence* or requiring an *adequate atrial communication*

- If there is limited/absent pulmonary blood flow (*ductal-dependence)*, this is a life-threatening problem and needs to be treated with prostaglandin

- If there is an inadequate *atrial communication* to support a viable circulation (e.g. tricuspid atresia, TGA, TAPVR), an atrial septostomy or immediate surgery is necessary.

Act III: Systemic outflow obstruction

- There are also several congenital heart diseases resulting in diminished *systemic* blood flow

- We will now look at two of them to dissect a few other important features of congenital heart disease

- Coarctation of the aorta
- Hypoplastic Left Heart Syndrome

Coarctation of the Aorta

- In fetal life there is very little flow across the distal aortic arch

- At times, this portion of the aorta fails to develop normally (e.g. mitral stenosis, aortic stenosis)

- There are 3 positions in the aorta for a coarctation to occur relative to the ductus arteriosus
  - Pre-, post, and juxta-ductal
  - The majority are *juxta-ductal*

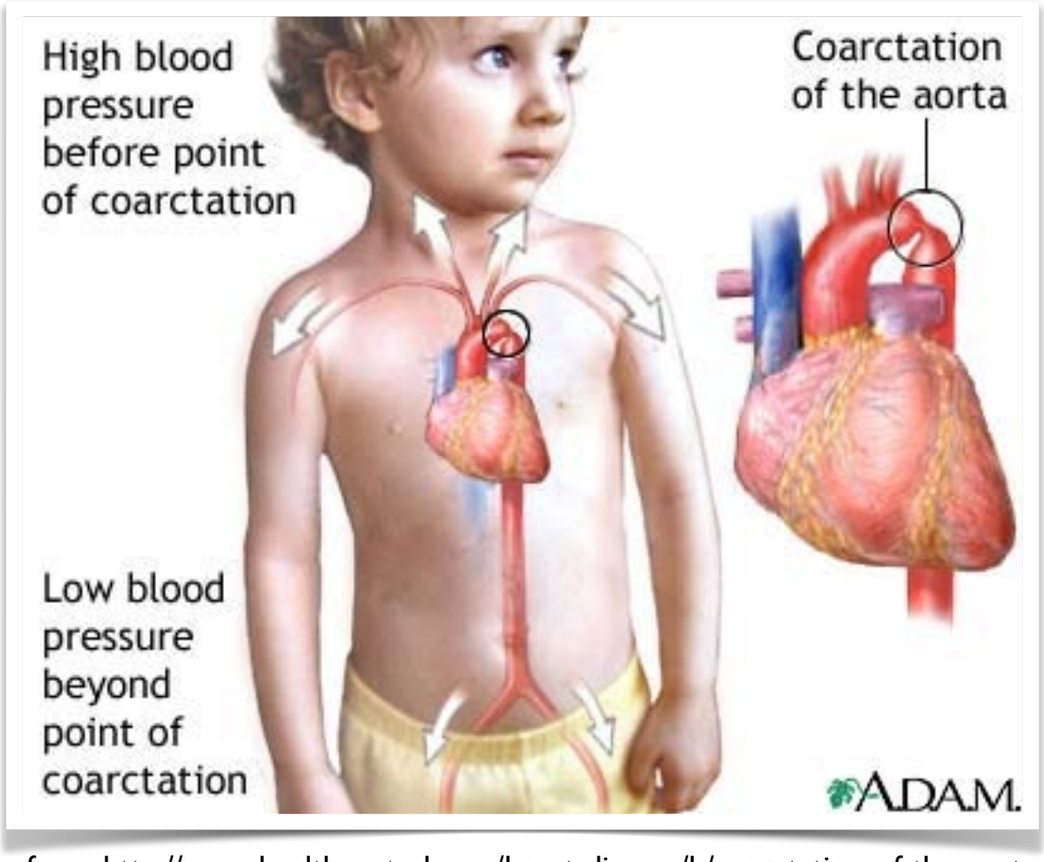
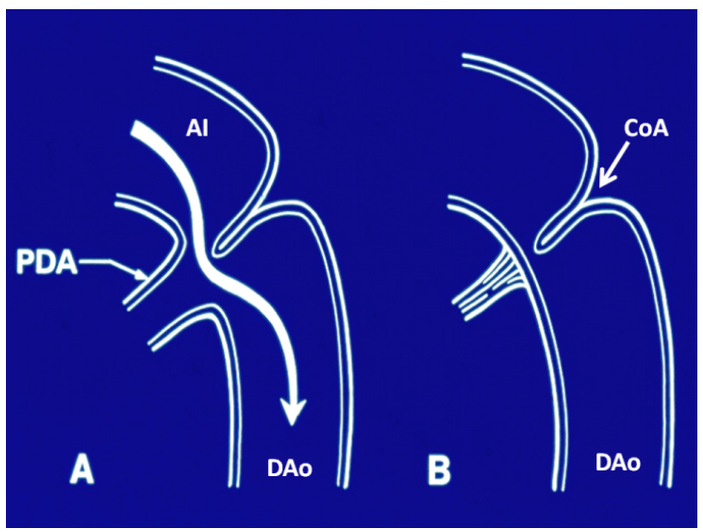


Image from: http://www.healthcentral.com/heart-disease/h/coarctation-of-the-aorta.html



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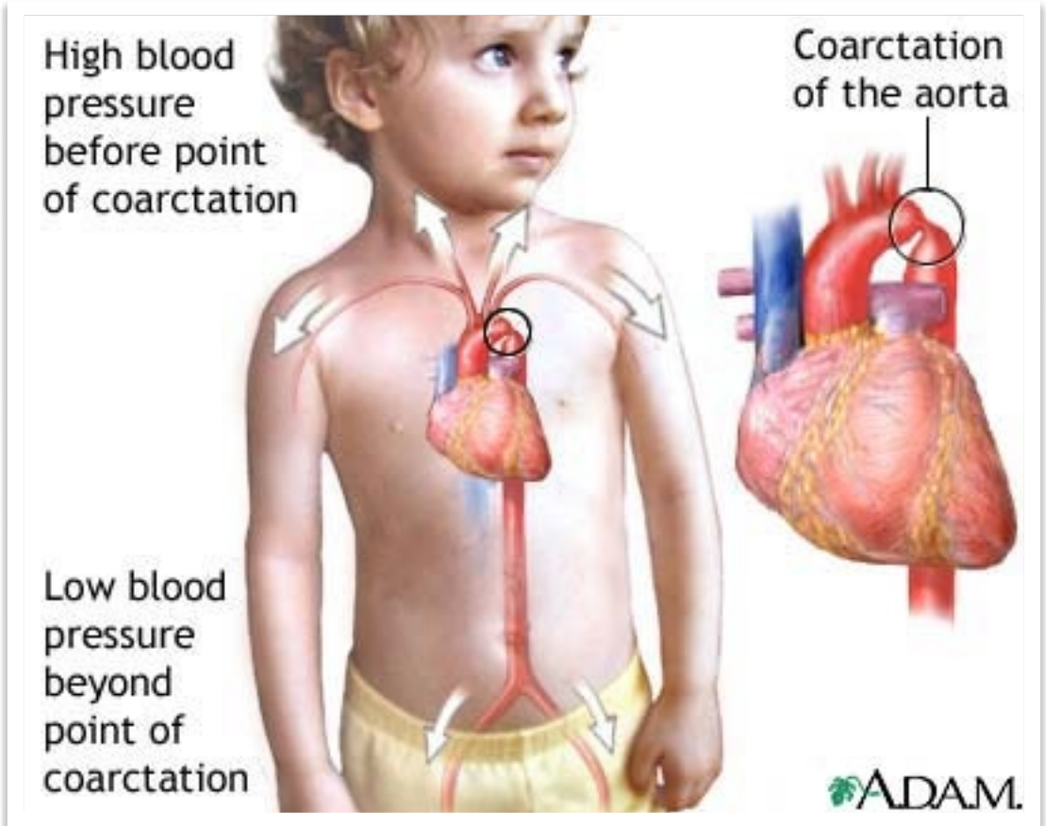
Doshi AR, Syamasundar Rao P (2012) Coarctation of Aorta-Management Options and Decision Making. Pediat Therapeut S5:006.

Coarctation of the Aorta



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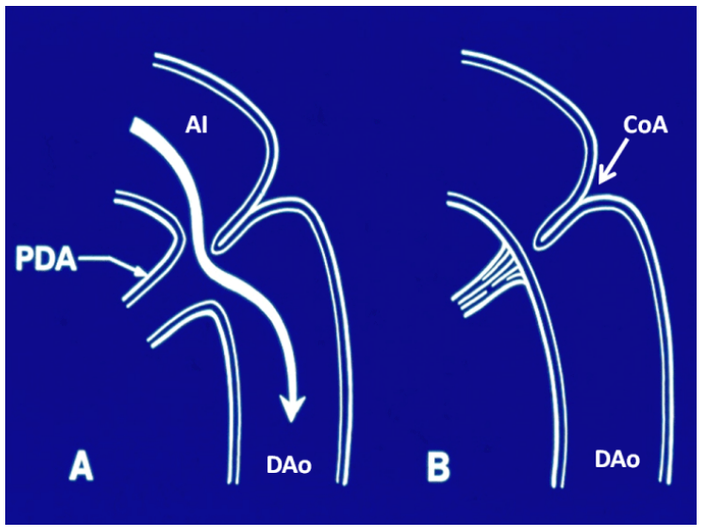

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- Many patients with coarctation will present during infancy as the PDA closes
- The closure of the PDA potentiates the obstruction to flow through that segment of the aorta
- If ductal patency can be maintained, flow into the aorta can occur via the PDA
- In this sense, we have a situation of *ductal dependent systemic blood flow!*

Coarctation of the Aorta

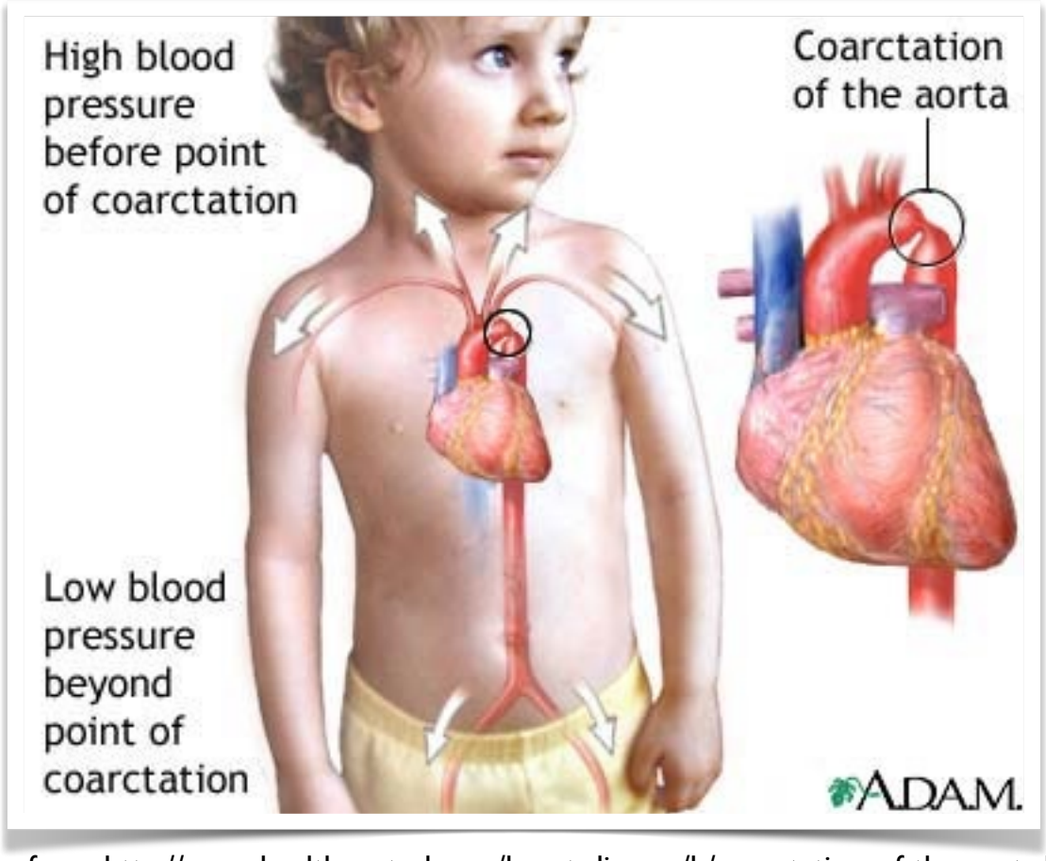
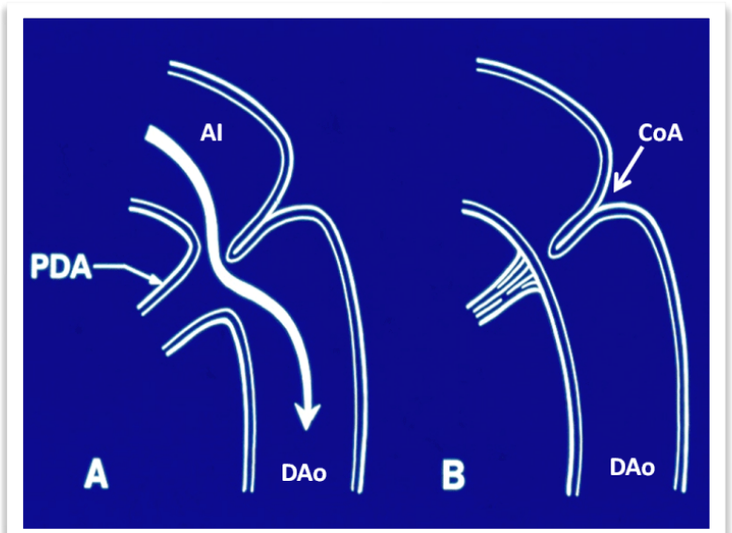


Image from: http://www.healthcentral.com/heart-disease/h/coarctation-of-the-aorta.html

- The hallmark physical findings in coarctation are *diminished femoral pulses* and *upper extremity hypertension*

- The critically ill infant may have signs of *congestive heart failure* as well as *hypoxemia in the lower half of the body.* Some will be in *shock!*
  - CHF results from back-up from obstruction
  - Hypoxemia results from right-to-left PDA flow

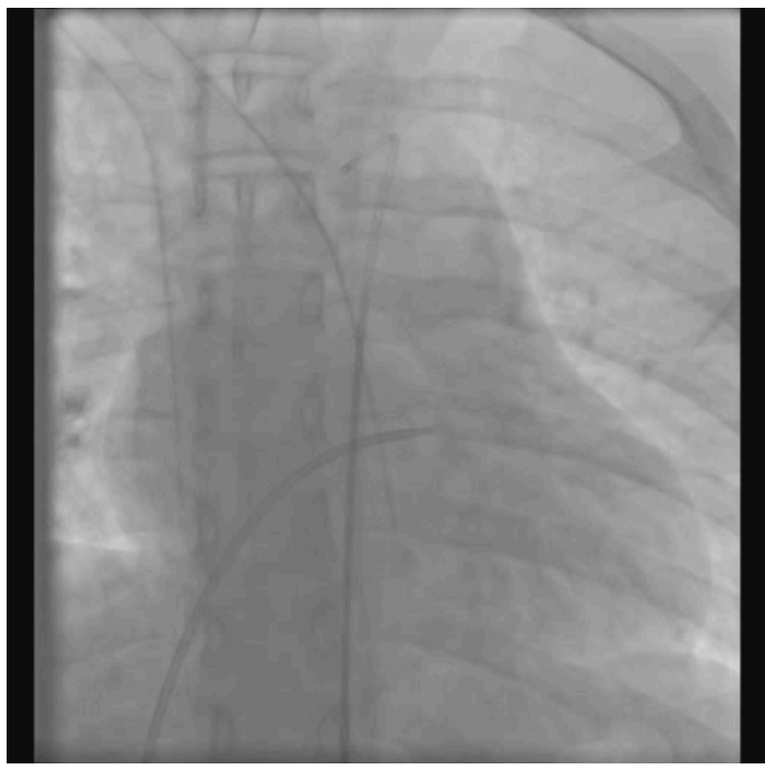


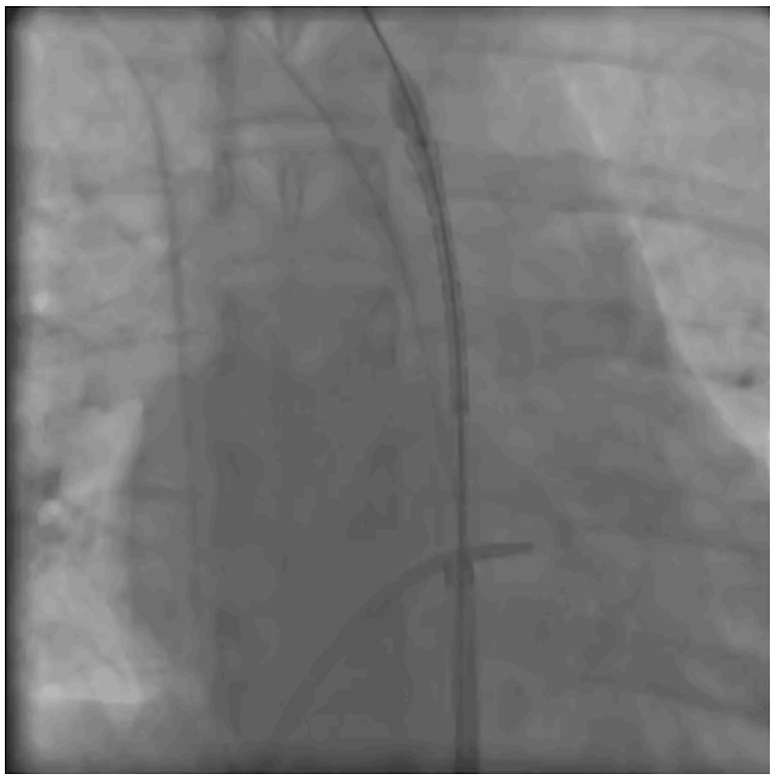
Doshi AR, Syamasundar Rao P (2012) Coarctation of Aorta-Management Options and Decision Making. Pediat Therapeut S5:006.

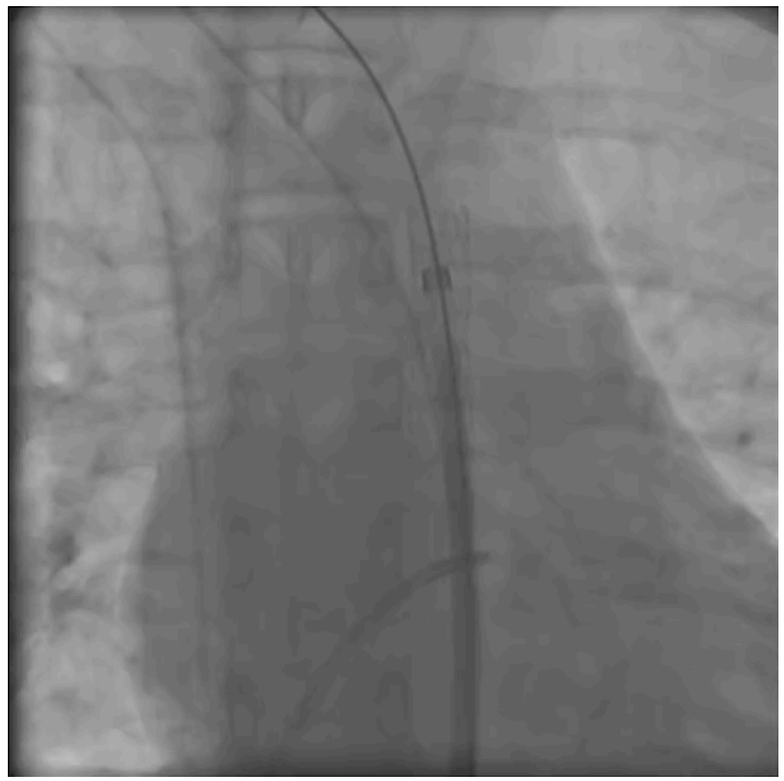
- Goal of management for the critically ill is to maintain the PDA, and immediately repair

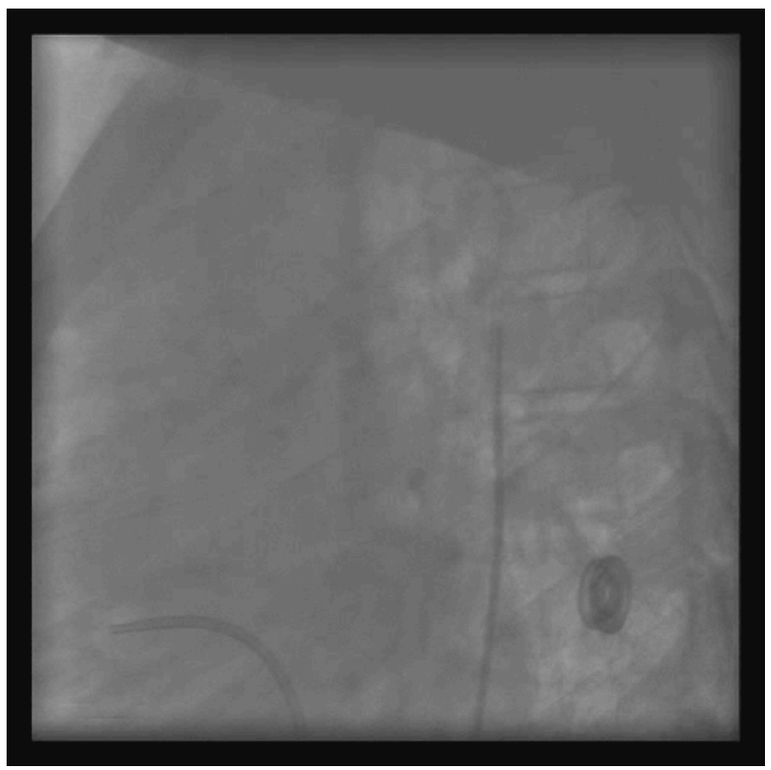
- Sometimes coarctation presents in an older child/ teenager who is found to be hypertensive

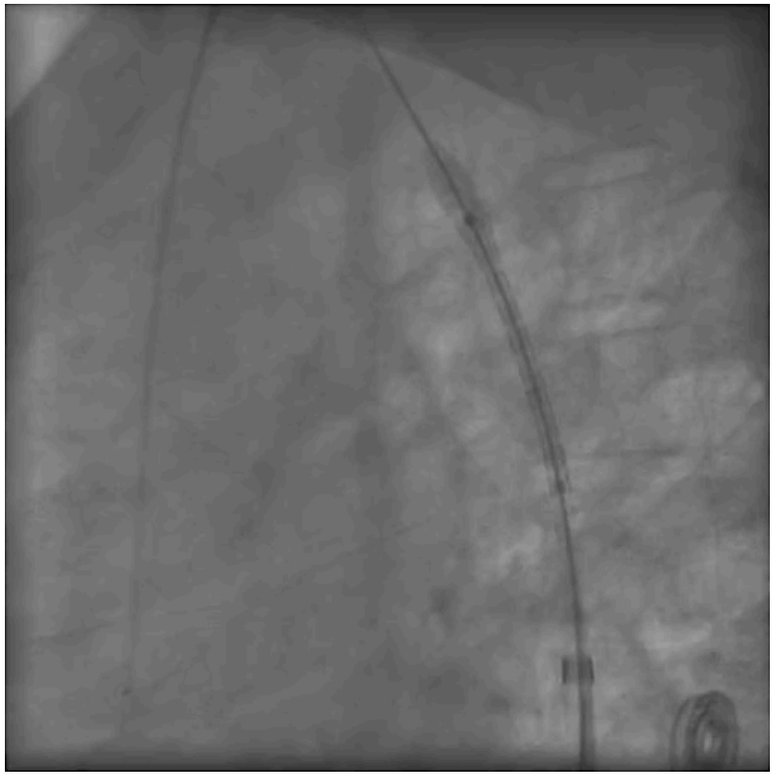
Coarctation of the Aorta

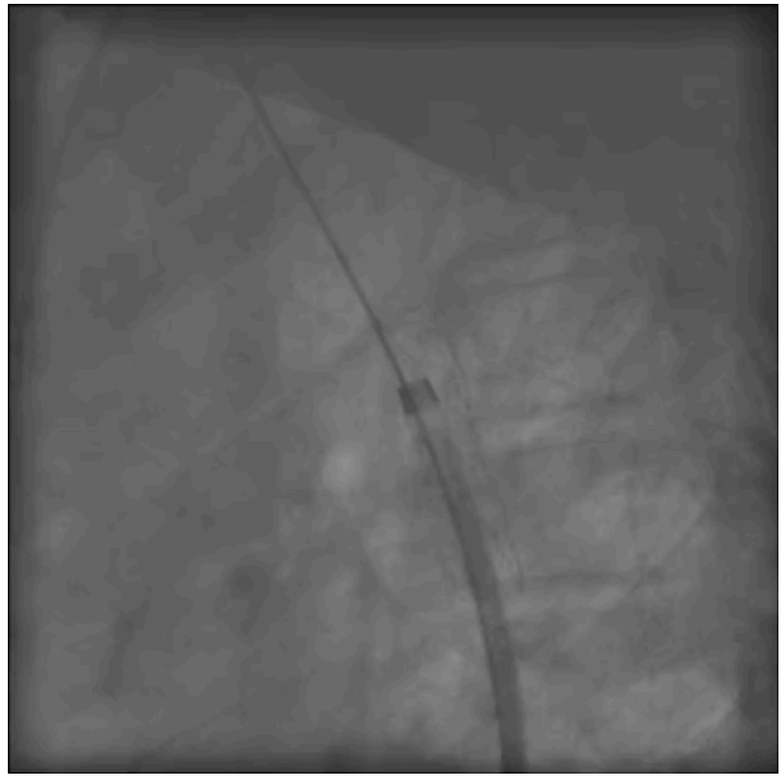












Hypoplastic Left Heart Syndrome (HLHS)

--another "single ventricle" lesion

- Combination of mitral stenosis/atresia and aortic stenosis/atresia

- Red blood must flow L-->R across a PFO/ASD
    - *what signs/symptoms would occur if the atrial communication is small?*

- Therefore *complete mixing* of blue and red blood

- Unlike tricuspid atresia, there is rarely a VSD in HLHS. The mixed blood goes out the pulmonary artery toward the lungs

- There's plenty of blood going to the lungs, but how does blood get out to the body?

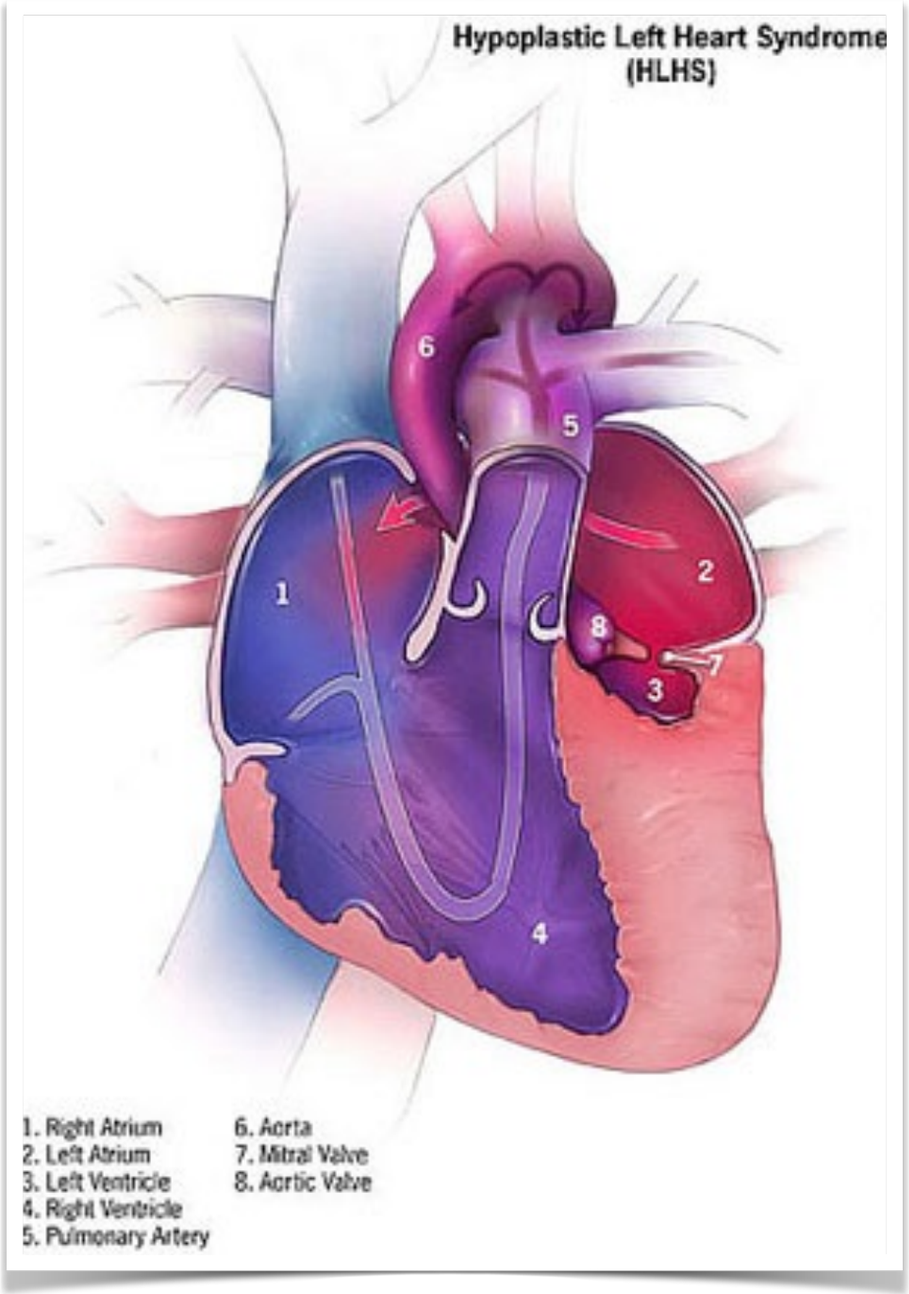


Image from: http://www.heartbirthdefect.com/heart-birth-defects/hypoplastic-left-heart-syndrome.php

Hypoplastic Left Heart Syndrome (HLHS)

--another "single ventricle" lesion

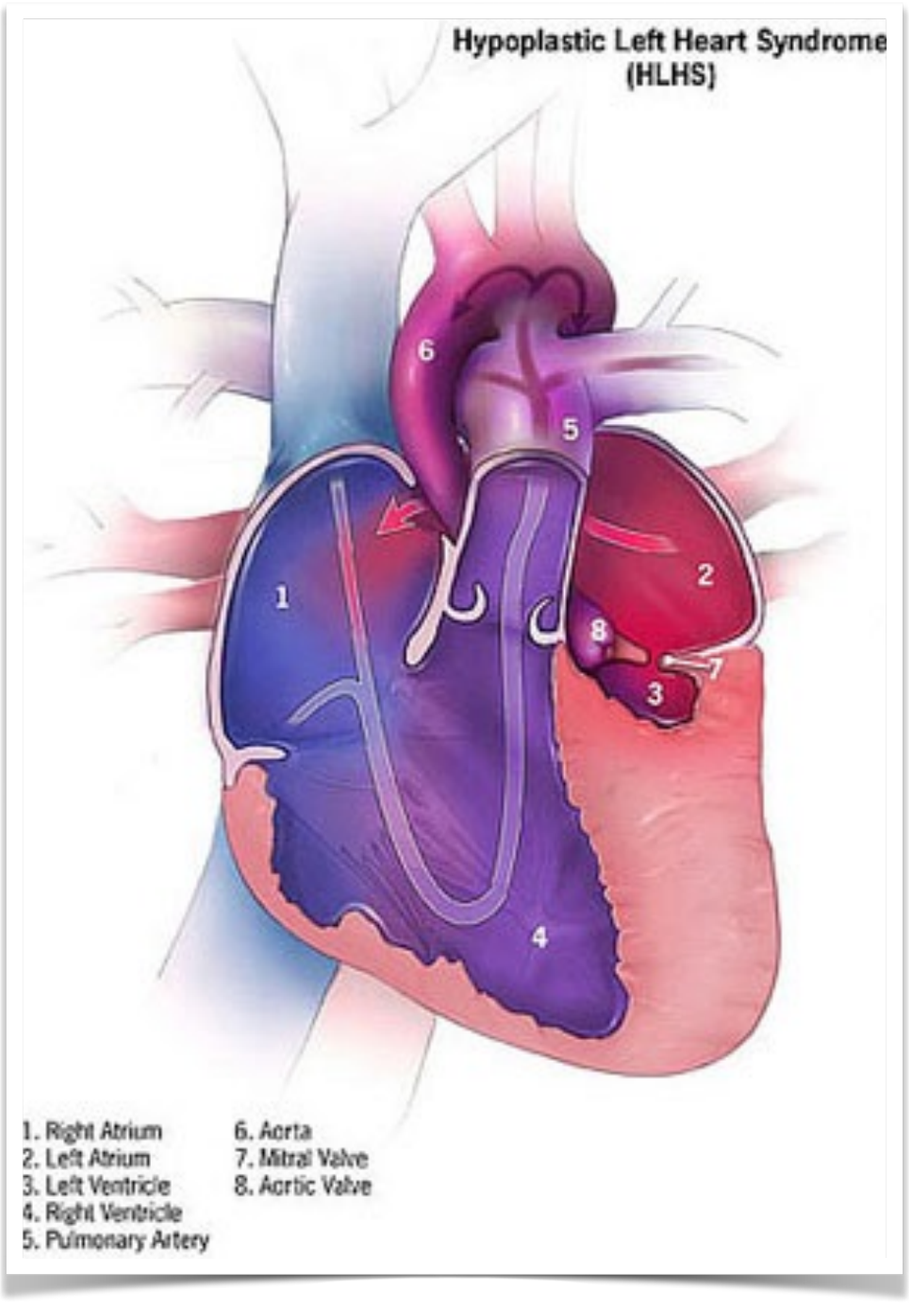


Image from: http://www.heartbirthdefect.com/heart-birth-defects/hypoplastic-left-heart-syndrome.php

This physiology is very similar to some of our other diseases with complete mixing of blue and red blood, but there are a few important features of HLHS:

1) Ductal patency is required for *systemic* blood flow

2) Resistance at the level of the atrial septum has slightly different consequences
   a. More profound hypoxemia
   b. Higher LA pressures-->CHF!

3) Coronary artery perfusion is tenuous

4) Single **right** ventricle!

Hypoplastic Left Heart Syndrome (HLHS)

--another "single ventricle" lesion

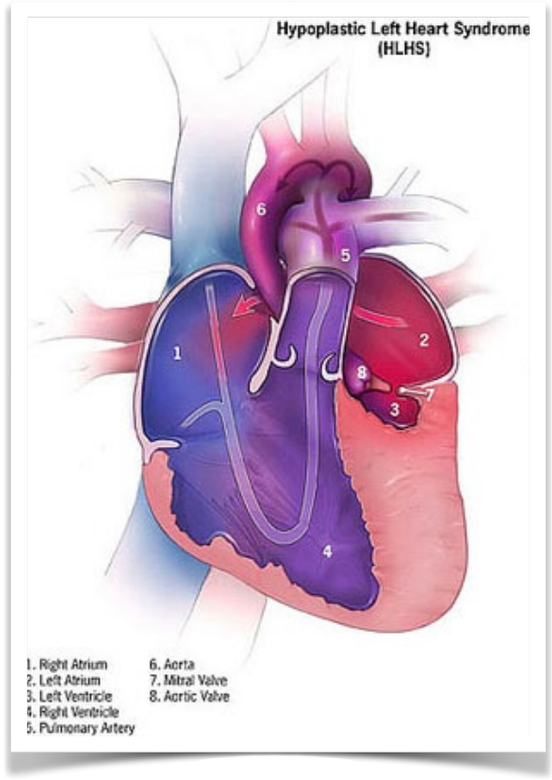


Image from: http://www.heartbirthdefect.com/heart-birth-defects/hypoplastic-left-heart-syndrome.php

What happens if the ductus closes?

What happens if the atrial septum closes?

Can we fix this?

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Act III Review: Systemic Outflow Obstruction

- Any obstruction to systemic outflow will create elevated pressures "upstream" of the obstruction

- *Elevated left atrial pressures result in CHF symptoms*

- If systemic outflow is completely obstructed, it needs to be supplemented by right-to-left flow at the PDA, resulting in *hypoxemia*

- The most extreme of these diseases is HLHS, where the LV does not form, and a single right ventricle pumps all blue and red blood to both vascular beds

Act IV: Repair/Treatment Options and Consequences

- First, we will discuss some general concepts of treatment

- Then, we will discuss some hemodynamic features of repaired cyanotic heart disease

(Remember, these patients will be showing up in your practice with greater frequency no matter what field of medicine you pursue!)

General Treatment Concept #1

- **Some congenital heart problems will spontaneously resolve**
  - Some ASDs
  - Some VSDs
  - Some PDAs

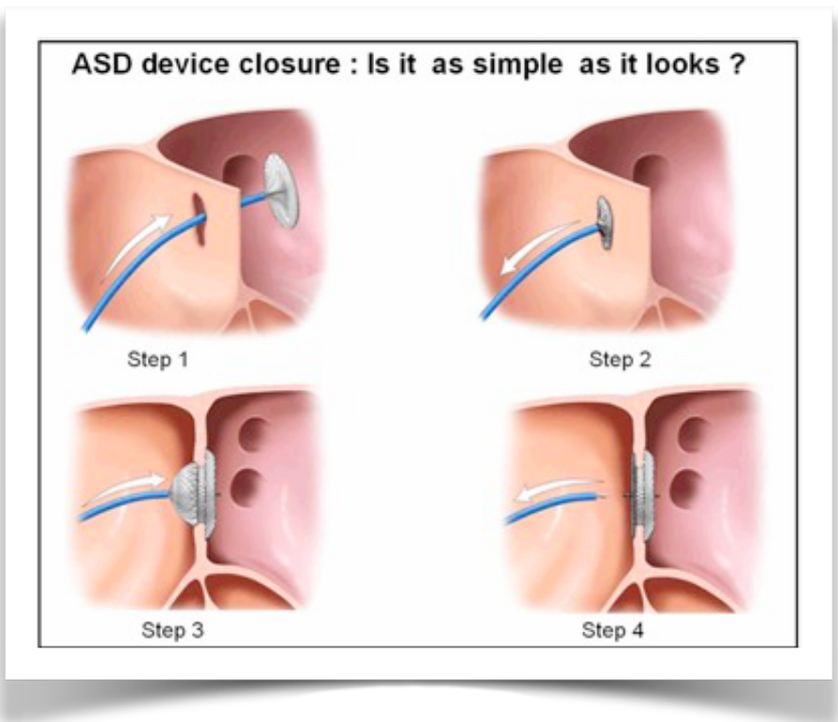
- These left-to-right shunts may require medical management of CHF symptoms until the defect gets smaller
  - Some VSDs, some PDAs

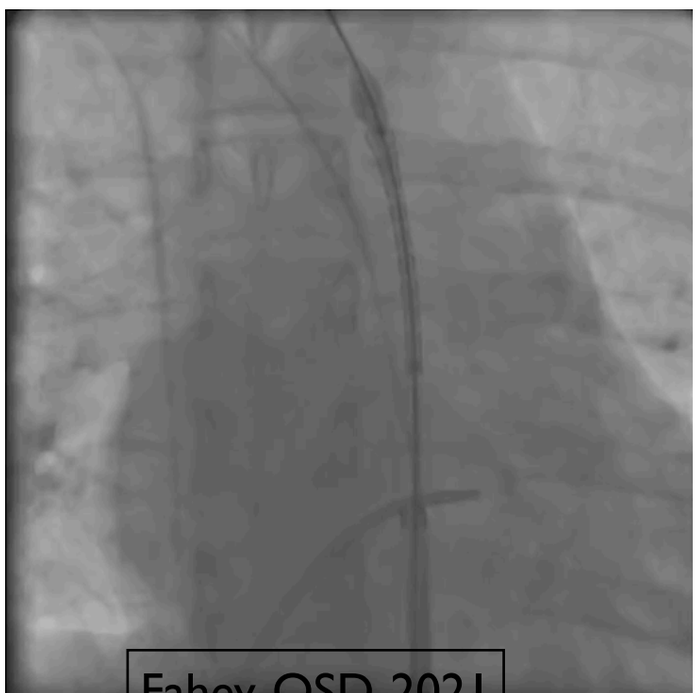
General Treatment Concept #1 (continued)

- While some of these lesions will spontaneously resolve, if they are not treated and a) cause enough pulmonary over-circulation or b) cause elevated left atrial pressure they will eventually result in *pulmonary vascular obstructive disease, a.k.a.* Eisenmenger's syndrome
  - We discussed this in Part 1

General Treatment Concept #2

- **When it comes to repair, there's more than one way to skin a cat**
- Holes/abnormal vessels can be closed with surgery or in the catheterization lab
- Small vessels/valves can be augmented with surgery or ballon dilated in the cath lab





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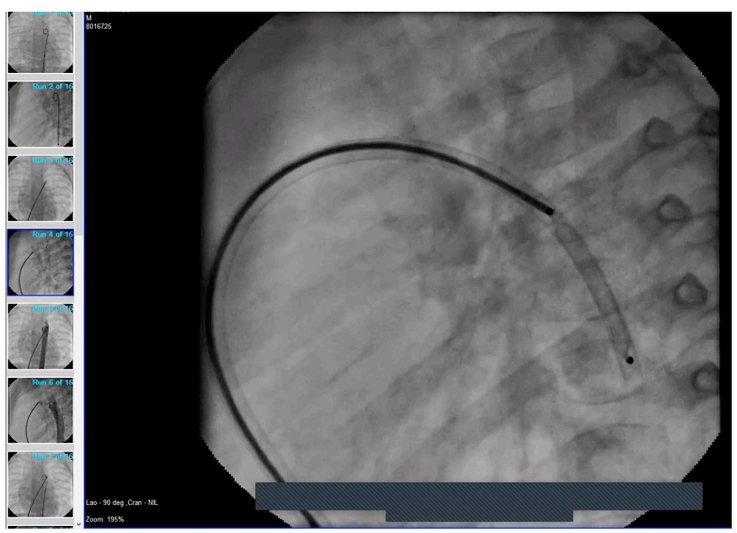

General Treatment Concept #3

Specific lesions require specific repair technique

Congenital Heart Condition	**Repair Technique**
ToF:	VSD closure, RVOT patch
Truncus arteriosus:	VSD closure, RV to PA conduit
TGA:	Arterial switch +/- VSD closure
TAPVR:	Anastomosis of PV confluence with LA, ligate vertical vein
Coarctation:	Coarctectomy, end-to-end anastomosis
Tricuspid Atresia/ HLHS:	Single ventricle palliation

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General Treatment Concept #3

Most cyanotic congenital heart lesions are not "fixed" entirely

Congenital Heart Condition	Residual Problems (Needed Procedures)
ToF:	-Chronic pulmonary regurgitation (Pulmonary valve replacement) -Residual branch pulmonary artery stenosis (balloon dilation/stenting of vessels)
Truncus arteriosus:	-RV-PA conduit stenosis (balloon dilation, surgical replacement) -Chronic conduit regurgitation (pulmonary valve replacement)
TGA:	-Branch pulmonary artery stenosis (balloon dilation/stenting) -Coronary artery ostial stenosis (surgery)
TAPVR:	-Pulmonary vein stenosis (balloon dilation/surgery)
Coarctation:	-Recoarctation (balloon dilation/stenting) -Systemic hypertension (medical therapy) -Increased risk of coronary artery disease (polypharmacy/surgery)
Tricuspid Atresia/ HLHS:	-"Failing Fontan" circulation (heart transplant)

General Treatment Concept #4

"Single Ventricle" lesions cannot be repaired, only palliated

- There is a *single ventricle* which must supply systemic blood flow
- Systemic venous return (blue blood) is re-routed to flow *passively* to the lungs
- This palliation takes place via *3 surgical stages*
  - Stage I (variable): Norwood or PA band or Blalock-Thomas-Taussig shunt
  - Stage II: Glenn shunt
  - Stage III: Fontan procedure

Single Ventricle Palliation: Stage I

Stage I aims at correcting the most threatening physiologic derangements (e.g. ductal-dependence, severe pulmonary overcirculation, inadequate ASD)

Problem	**Solution**
Ductal-dependent pulmonary blood flow	Blalock-Thomas-Taussig Shunt
Pulmonary overcirculation	Pulmonary Artery Band
Inadequate ASD	Atrial Septectomy
Severe aortic hypoplasia	Norwood Procedure

Single Ventricle Palliation: Stage I

Blalock-Thomas-Taussig Shunt

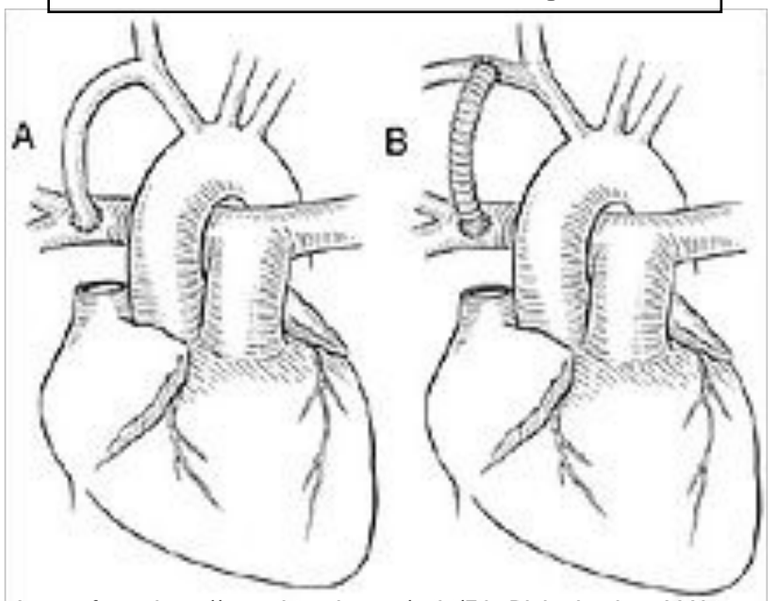


Image from: http://en.wikipedia.org/wiki/File:Blalock_shuntWiki.jpg

Pulmonary Artery Banding

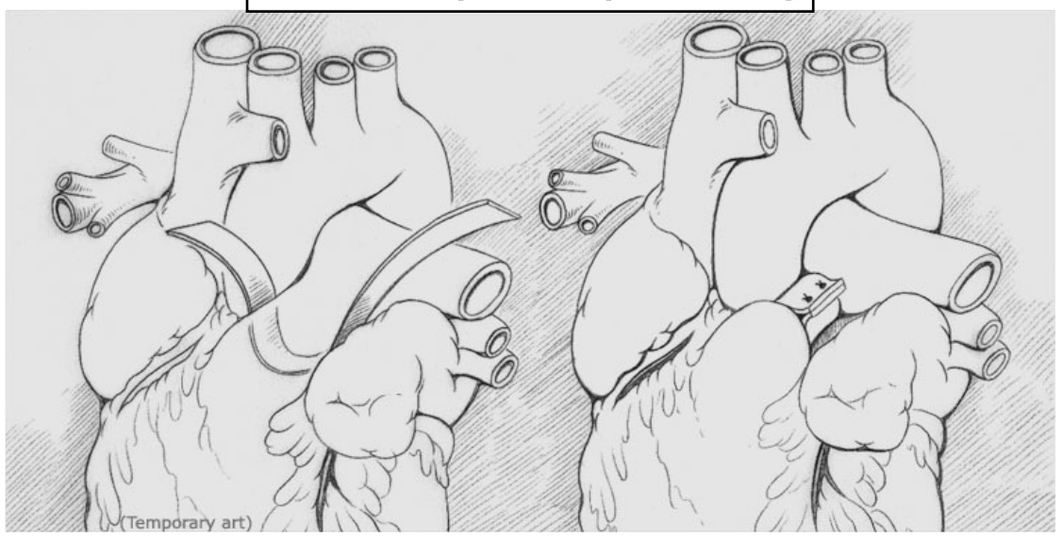


Image from: http://www.yale.edu/imaging/chd/surgery_pa_banding/index.html

Blalock-Hanlon Septectomy

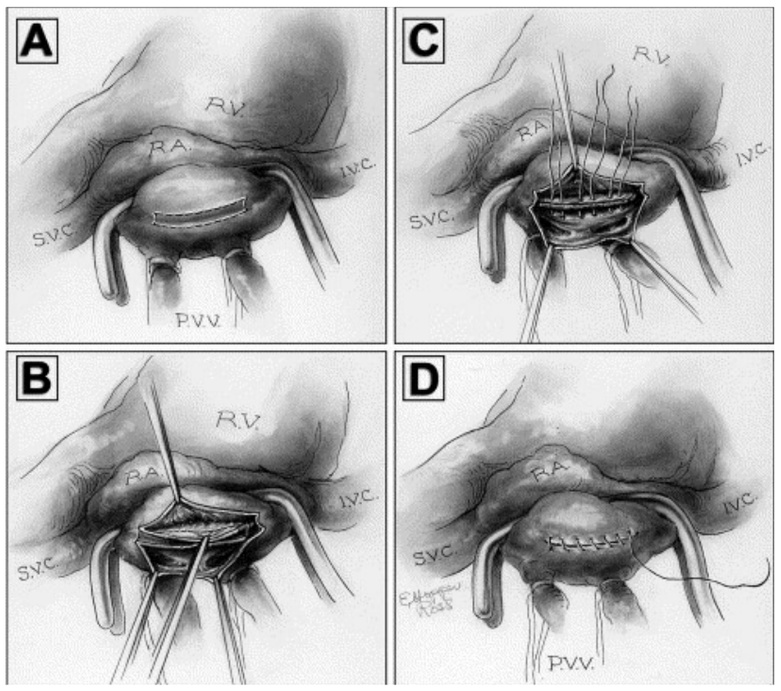
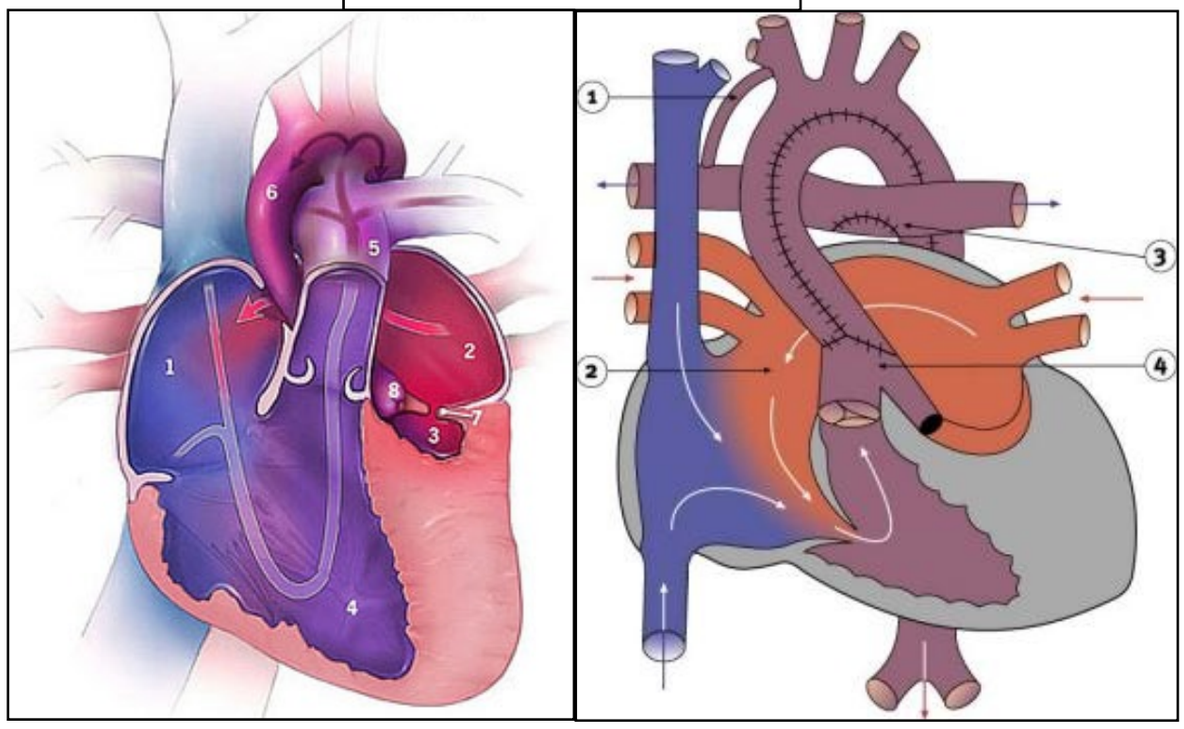


Image from: Ann Thorac Surg Volume 77, Issue 6, June 2004, Pages 2250–2258

Norwood Procedure



Images from: http://www.heartbirthdefect.com/heart-birth-defects/hypoplastic-left-heart-syndrome.php and http://iheartmoses.com/wp-content/uploads/2012/06/2_21.jpg

Blalock-Thomas-Taussig Shunt

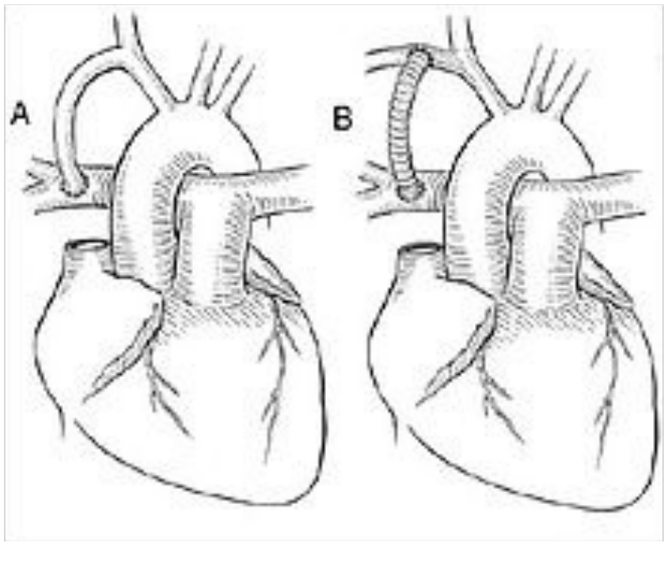


Image from: http://en.wikipedia.org/wiki/File:Blalock_shuntWiki.jpg

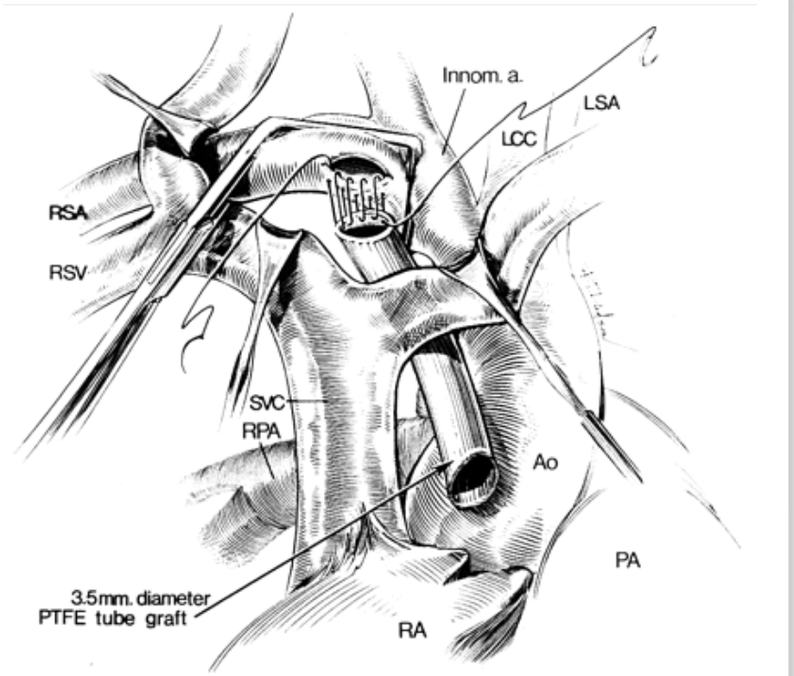
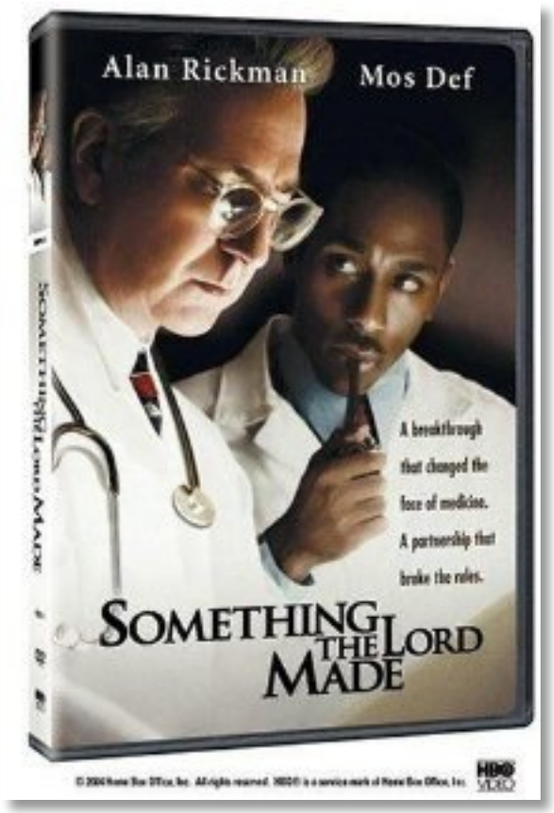


Image From: Circulation. 1995; 92: 256-261

The story of the BTTS is fascinating.

We don't have time to explore it now...




Sidetracks Blood and Oxygen to Otherwise Starved Lungs

HEAD
LUNGS
HEART

The "Blue" Babies' Blood Lacks Vital Oxygen Because the Artery (1) From the Heart to the Lung Is Constricted. By Severing an Artery of the Arm (2), Tying It Off (3) and Attaching It to the Lung Artery (4) the Constriction Is By-Passed.

Saving Our Doomed 'Blue' Babies

By Robert D. Potter
Science Editor

A WOMAN physician's courageous research and imagination, and the skill of one of the world's great surgeons have combined to bring hope that many "blue" babies hitherto considered doomed to early death—may be saved.

These babies are blue because they are suffering from a lack of oxygen in their blood streams, in a condition known as cyanosis. The artery from their heart to their lungs is so constricted that their blood never gets oxygen to make cheeks rosy.

Their lips are blue. Their toes are blue, and they can walk only a few feet without exhaustion. Doctors used to give them only a few tortured years to live.

But now medicine can give hope . . . and more . . . for since Nov. 20, 1944, Dr. Blalock, Professor of Surgery at Johns Hopkins University in Baltimore, has been conquering the "blue" baby malady by rerouting an artery from the arm and making it carry blood to the lungs where it can receive its vital oxygen.

Nearly 70 operations have been performed on "blue" babies. In many cases almost miraculous recovery has come.

It is Dr. Blalock's fingers that wield the knife in the delicate operation that exposes the heart and transplants its vital arteries. But behind the brilliant operation he has perfected are years of painstaking research by Dr. Helen B. Taussig.

Daughter of the late Prof. F. W. Taussig, world-famous Harvard economist, Dr. Taussig had watched "blue" babies come to her heart clinic at Johns Hopkins Hospital.

In many cases she discovered that the artery leading to the lung from the heart was narrowed so that an insufficient supply of blood was reaching the lungs to receive its vital oxygen. Dr. Taussig reasoned that a surgical operation might be able to short-circuit the constriction and sidetrack blood into the lungs. On the blood would pick up its life-giving oxygen. Then it would go back to the heart again to move outward through the body.

But could it be done? It is one thing to have a plumber re-arrange a piping system and something quite different to lay bare the human heart, sever one of its main arteries, splice it to another main artery and sustain life in the patient in the process, Dr. Blalock said he would try.

Since the pioneer attempt the operation has been largely successful, although it is one filled with danger. Among the first 70 patients, 14 died. The odds are 5 to 1 for success.

Now that the news of Dr. Blalock's operation is known through the country the list of patients grows daily. Blue little Bonnie Stewart of Florida, daughter of a daddy killed on Saipan, went to Baltimore with her grandmother. Today Bonnie walks and plays like other children.

The case of six-year-old Mike Schirmer—the boy with the "itchy zipper"—shows what can be done.

Six-Year-Old Mike Schirmer of Baltimore Could Walk Only Five Feet Without Resting Before the Operation. He Shows His "Itchy Zipper"—the Incision for the Operation.

Little Bonnie Stewart of Florida Is Another of the 70 Children Saved by the New Johns Hopkins Surgery.

Mike's "itchy zipper" is the healing incision over his heart where Dr. Blalock went in to do the operation. But let his mother tell his story:

"Michael could only walk five feet and then he'd have to squat down on the sidewalk and rest.

"I had to wheel him everywhere.

was no hope that Mike could grow up. But then came new hope, for Dr. Mansiederfer told us about the operation of Dr. Blalock. "They took him to the operating room and brought him back two hours later. It was a miracle.

"After only two weeks of convalescence he came home and he has been on the go ever since. If anyone wants anything he'll run and get it. He's up and down stairs 75 times a day. He climbs on bureaus and tables just for the joy of jumping off. He wears me out. But I love it."

The Blalock-Taussig operation is

branches of the pulmonary artery (to the lungs) are two large blood vessels. One connects the heart and the arm, the other the heart and the head. Dr. Blalock chooses the most convenient—usually the arm artery—and severs it. One end is clamped off and the other closed permanently.

The end nearest the heart is then spliced to the nearest branch of the pulmonary artery. The clamps are removed and the blood that would ordinarily flow to the arm goes into the lung. There it becomes enriched with vital oxygen and the baby's blue lips quickly begin to turn red.

What happens to the arm?

Nature has provided other blood

From: http://www.medicalarchives.jhmi.edu/amwklg.jpg

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Single Ventricle Palliation: Overview

"Stage I" surgery addresses the most critical aspects of the structural heart defect, whatever those may be

Following Stage I, all single ventricles will undergo the same second stage (Glenn) and third stage (Fontan) surgeries

The second two stages aim to re-route systemic venous blood directly to the lungs

Single Ventricle Palliation: Stage II--Glenn Shunt

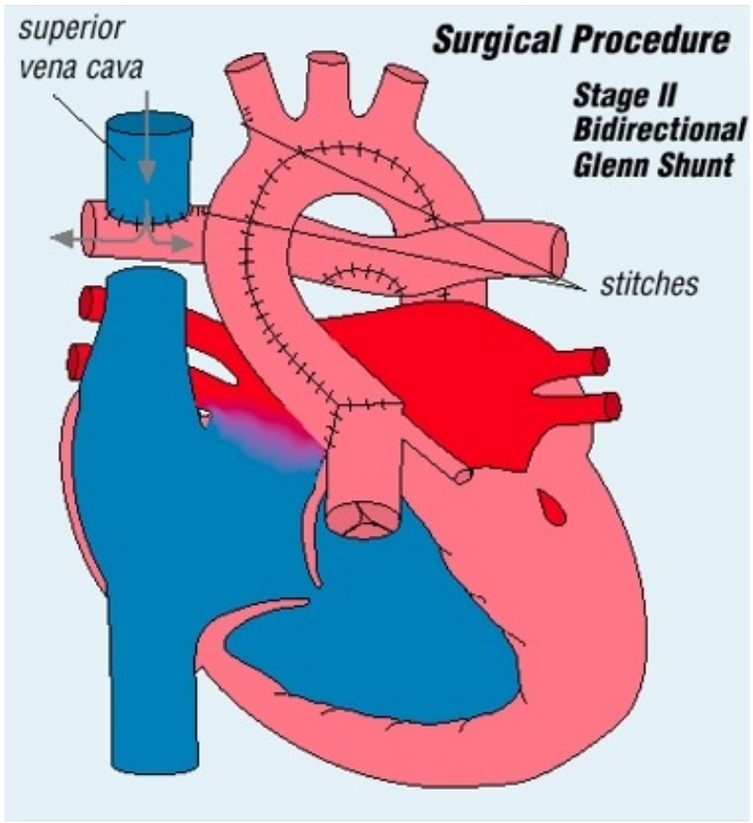
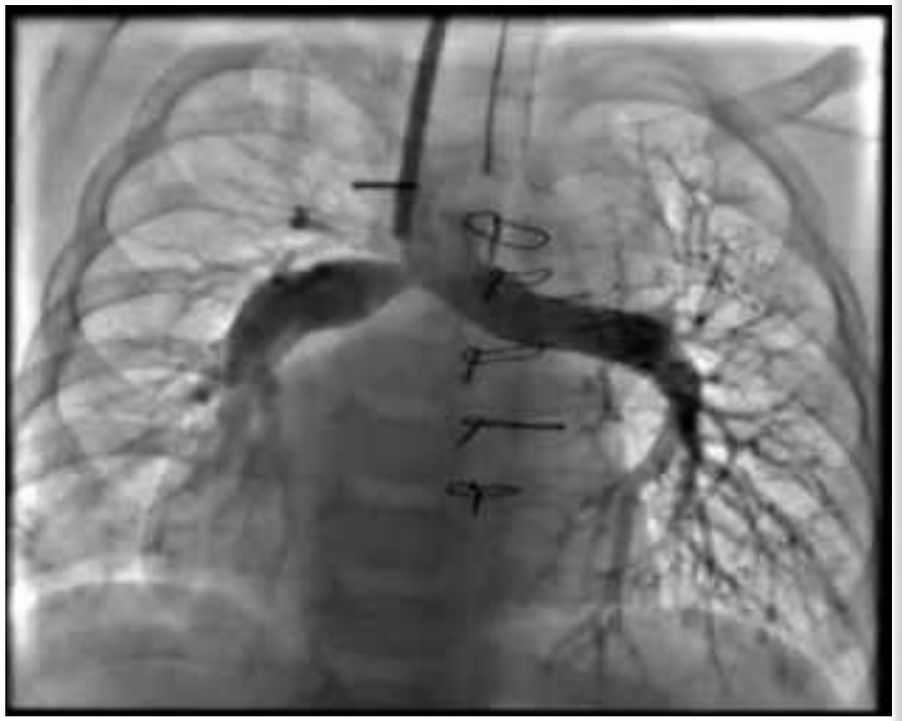


Image from: http://www.chitexas.org/images/glenn.jpg



https://www.youtube.com/watch?v=PuDSd0JPzzE

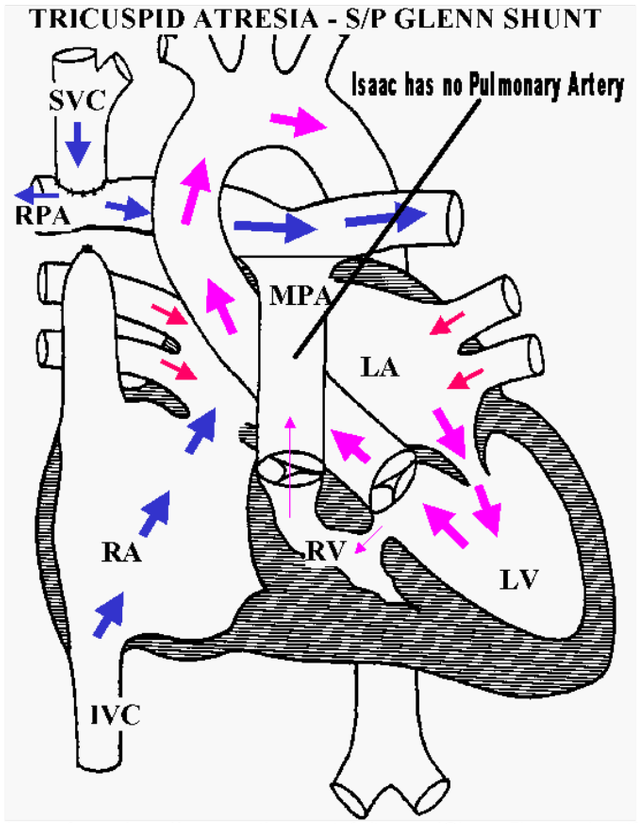


Image from: http://www.smileyman.info/images/Glenn_Flow_color.gif

The Glenn re-routes blood from the upper half of the body directly to the lungs.

Single Ventricle Palliation: Stage III--the Fontan

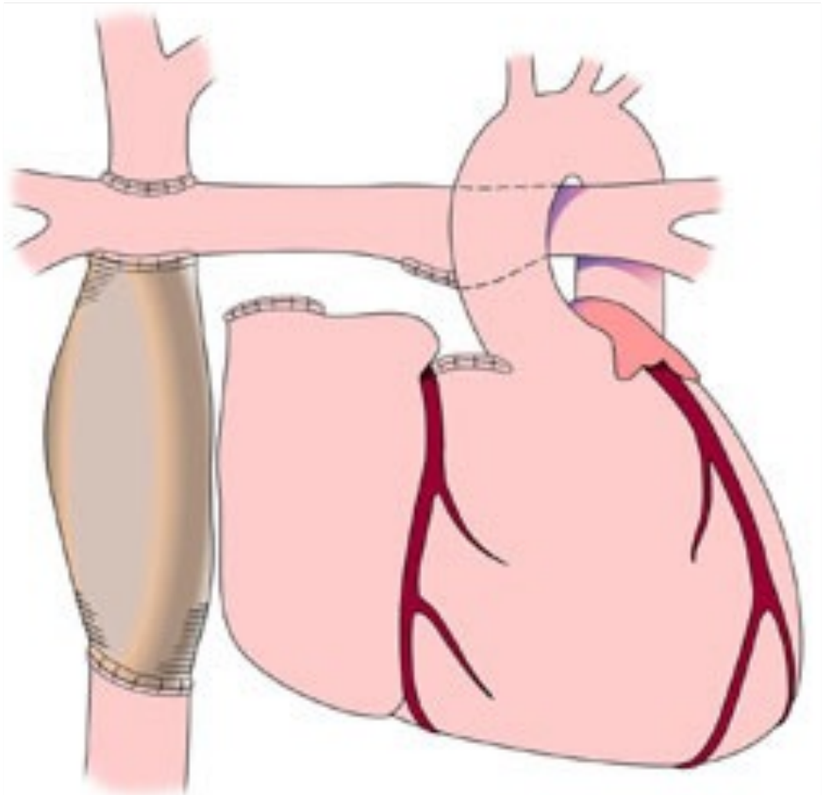


Image from: http://www.childrenshospital.org/cfapps/mml/viewBLOB.cfm?MEDIA_ID=1839

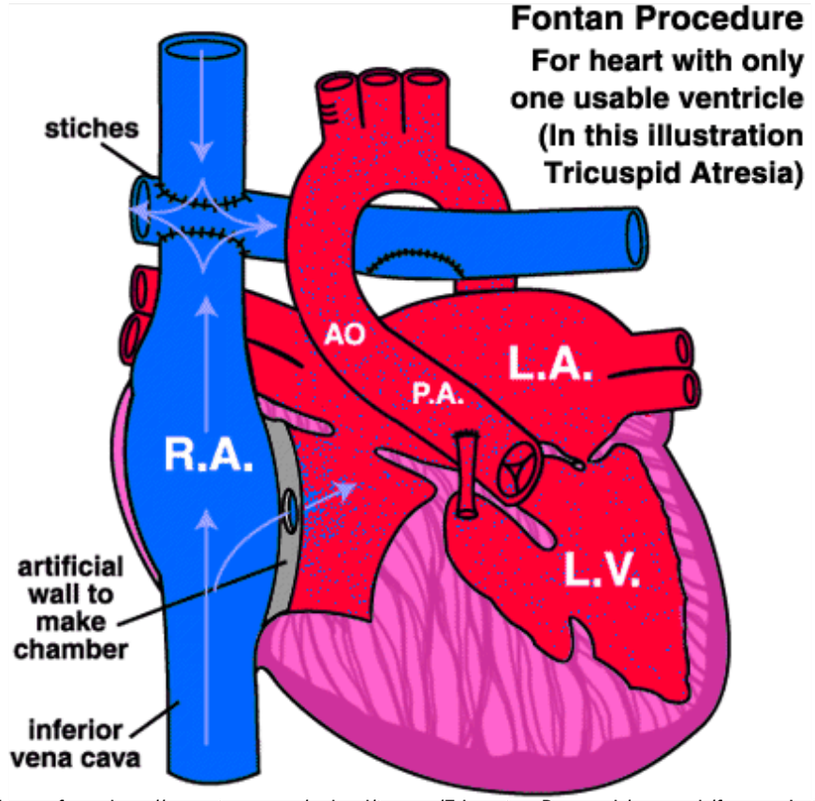
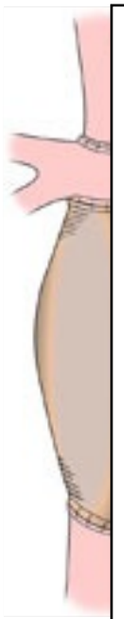


Image from: http://www.inova.org/upload/images/Education-Research/research/fontan_1.gif

The Fontan re-routes blood from the lower half of the body directly to the lungs.

What is the consequence of passive blood flow to the lungs?

Single Ventricle Palliation: Stage III--the Fontan



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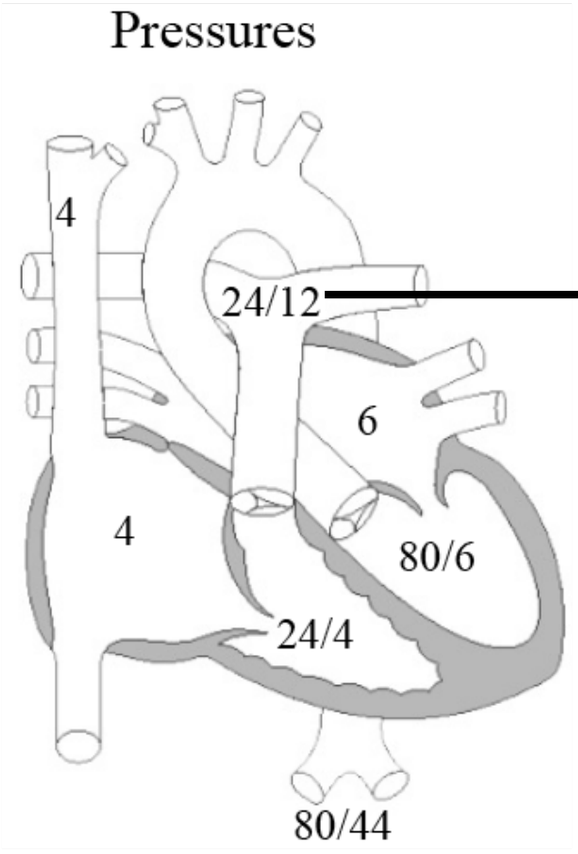
The Fontan re-routes blood from the lower half of the body <u>directly to the lungs</u>.

What is the consequence of passive blood flow to the lungs?



Mean PA pressure = 16mmHg

What is the central venous pressure in a Fontan?

ΔP = Q•R

In other words, pressure is a *byproduct* of flow and resistance

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Single Ventricle Palliation: Stage III--the Fontan

- What is the consequence of such high CVP?
  - hepatosplenomegaly
  - lower extremity edema
  - ascites
  - pleural effusions

Ascites and pleural effusions don't drain well in a Fontan. Why not?

What effect will a pleural effusion have on the pulmonary vascular resistance? On Fontan pressure?

Act IV: Repair/Treatment Options Review

- Some defects close spontaneously
- Others require surgical or catheter intervention
- Many require medical therapy as well
- Invasive procedures are tailored to the underlying anatomy
  - These procedures often leave significant long-term morbidities in their wake (e.g. tetralogy of Fallot repair)

Act 4: Repair/Treatment Options Review

- Single ventricle lesions can only be palliated
- Single ventricle palliation restores normal cardiac output and normal oxygen saturation at the expense of very elevated CVP and elevated afterload on the remaining ventricle
- Finally, it's very important to understand that kids with cyanotic congenital heart disease grow up to be adults with ongoing cardiac issues

  —rarely is there a true "fix" to these complicated problems

  —therefore, you will need to be aware of these problems and make sure your patients are properly cared for!

Thank you!!!

Questions? Email me:

michaelc.fahey@umassmemorial.org