

Transposition of the Great Arteries (TGA)

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Agenda

- Background of TGA
 - Epidemiology
 - Anatomy
 - Clinical presentation
- Meet Baby L: Differential diagnosis
- Menu of Radiologic Tests for TGA
 - Plain film
 - Echocardiography
 - Cineangiography
- Treatment and complications of TGA

Background of Transposition of the Great Arteries (TGA)

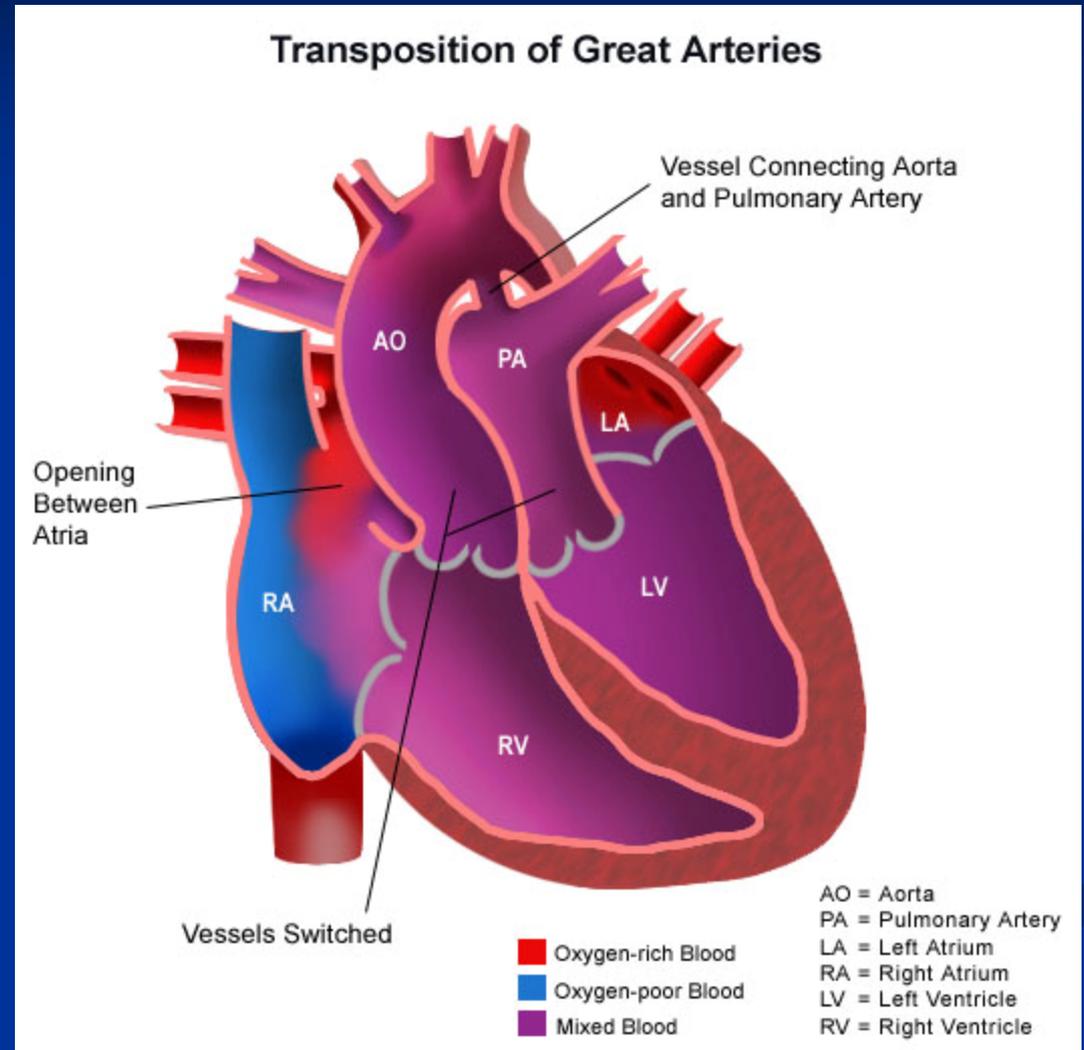
- Simplified definition: aorta comes off of the RV, pulmonary artery comes off of the LV



- Many anatomic varieties, most common is called D-transposition (often called Complete TGA). This is the form we will discuss in this presentation.
- Most common cyanotic heart defect recognized in the neonate
- Approx 8% of all congenital heart disease cases. 1 in 4,000 live births.
- Related to embryologic failure of common arterial trunk to spiral and septate normally
- Without treatment, 90% infants die by 1 yr
- 5-year survival rate after surgery is more than 80%

Anatomy of TGA

- Concordant atrioventricular connection, but ventriculoarterial discordance → 2 parallel circulations
- Associated abnormalities: VSD in approx 50%, PS. Less common: ASD, PDA, PFO.
- Incompatible with life unless mixing of circulations via an ASD, PFO, VSD, or PDA. Less mixing → more profound hypoxia.



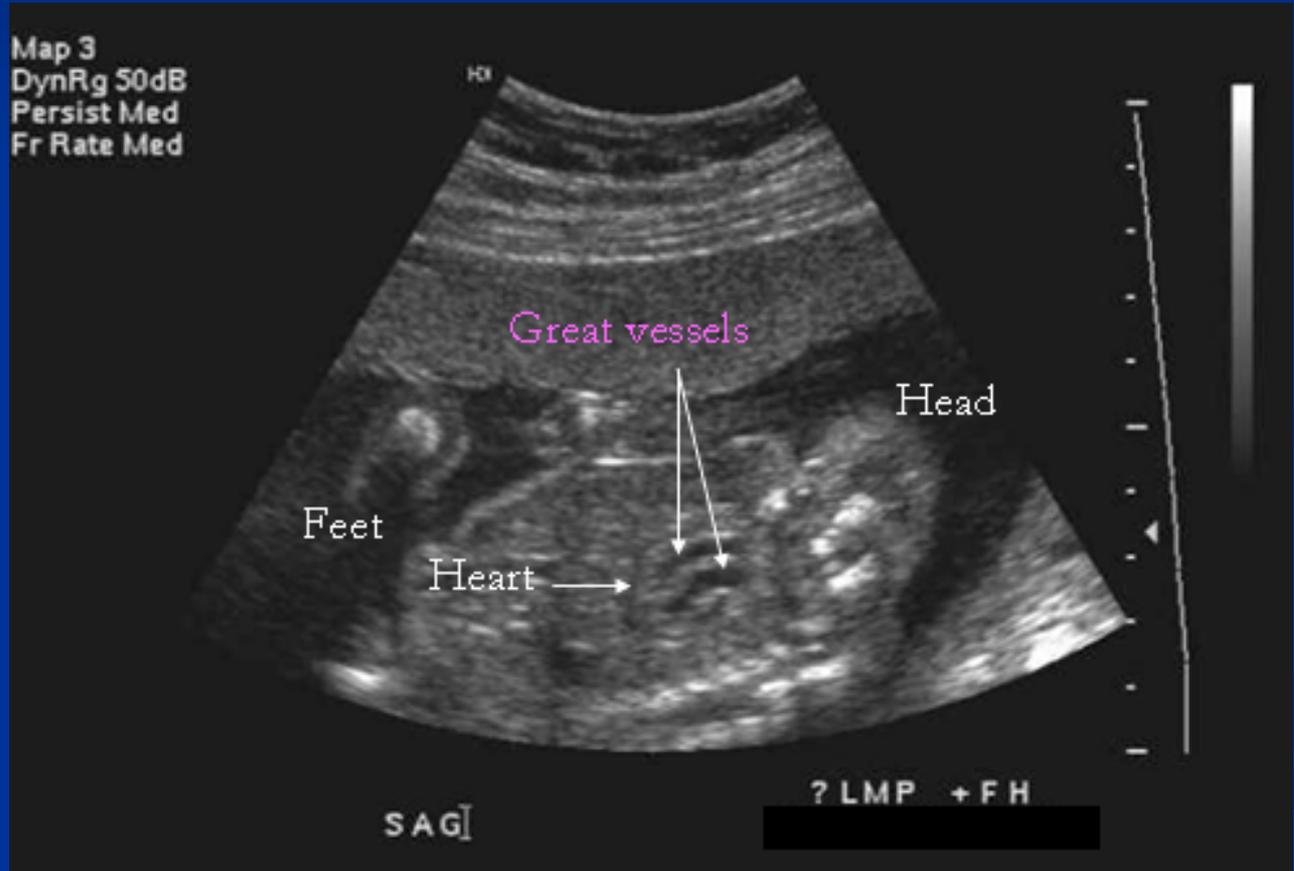
Typical Presentation of TGA

- Clinical presentation:
 - If the infant has an intact ventricular septum: cyanosis at birth (at least by 48hrs because by then the ductus arteriosus has closed), often acidosis.
 - If the infant has a large VSD: less severe cyanosis, but CHF from left ventricular volume overload
- PE:
 - tachypnea (b/c hypoxic), tachycardia
 - no murmur unless other lesions present
 - palpable right ventricular impulse since RV faces systemic pressures
 - accentuated S₂ due to aortic valve closure located anterior, just under chest wall

Prenatal Diagnosis

Companion Patient #1: TGA on Fetal Ultrasound

- Most common way TGA is diagnosed in Boston
- Key: see **two parallel great vessels**
- Normally, the vessels should cross each other



Courtesy Dr. Rola Shaheen, BIDMC

Our Patient Baby L: Presentation

- Male infant in first day of life transferred from outside hospital with low oxygen saturation
- No significant prenatal complications or findings
- Born at 42 3/7 weeks gestation, BW 3560 grams, Apgars 9 and 9
- “Dusky” after delivery
- Sats in low-70s on room air, so placed on oxyhood
- Sats in low-80s despite 100% FiO₂, so initiated HFOV (high frequency oscillation ventilation) and nitric oxide, and transferred from outside hospital

Differential Diagnosis of Neonatal Respiratory Distress

1. Aspiration of meconium or amniotic fluid
2. Congenital heart disease, especially cyanotic
3. Diaphragmatic hernia
4. Hyaline membrane disease/bronchopulmonary dysplasia
5. Pneumonia
6. Pulmonary immaturity
7. Respirator therapy (PEEP)
8. Transient tachypnea of the newborn

Menu of Radiologic Tests for Evaluation of Congenital Heart Disease

- Plain film
- Echocardiogram
- Cineangiogram
- For complications: CT and MR

Plain Film Imaging

Advantages:

- Non-invasive, fast, relatively inexpensive, easily accessible
- Regarding TGA: Identify abnormalities that point against TGA (eg. massive cardiomegaly, bony abnormalities)

Limitations:

- Nonspecific – seldom indicates specific cardiac anomaly, cannot visualize internal heart structures
- Regarding TGA: plain film findings are variable, and not related to degree of cyanosis!

TGA on Plain Film: Introduction

- TGA may have a variable appearance on CXR depending on presence and size of shunts, age of infant, co-existing conditions, and more.
- Let's continue to view 3 different patients with TGA on CXR before viewing Baby L's CXR.

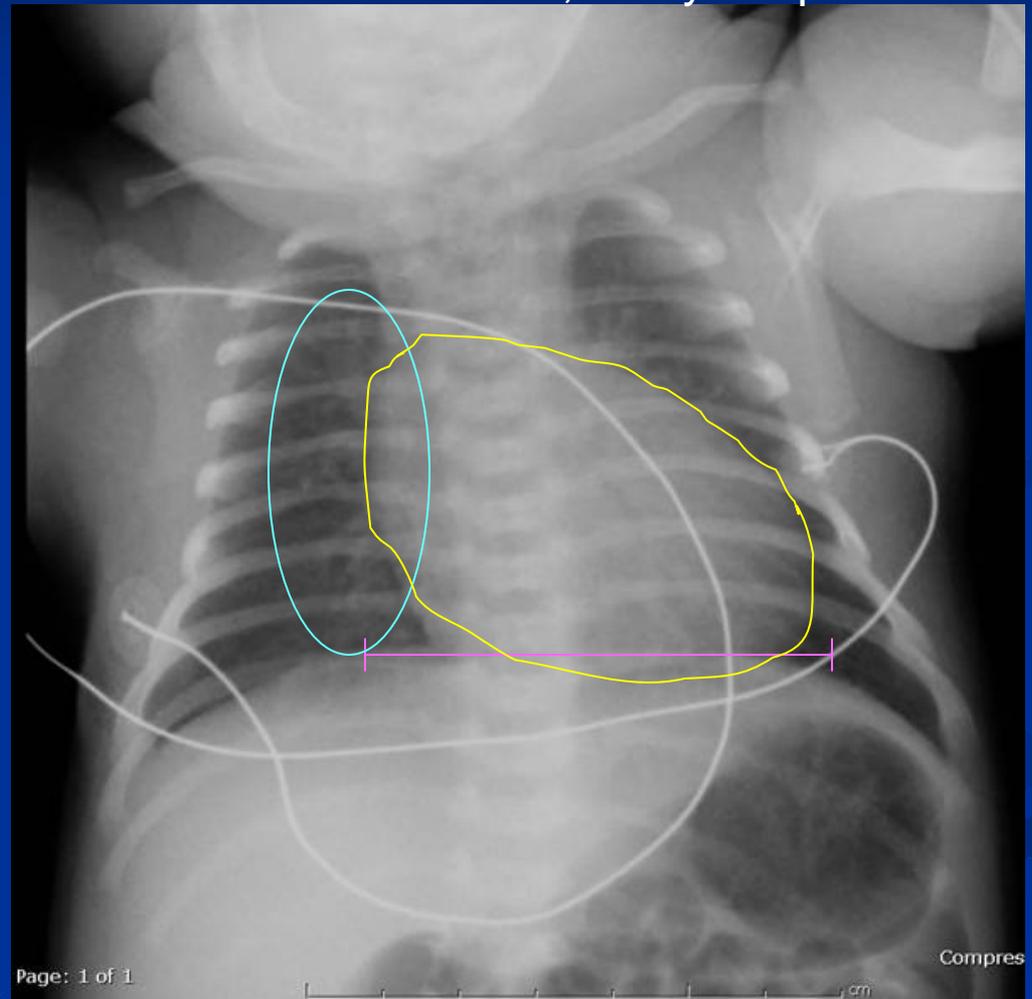
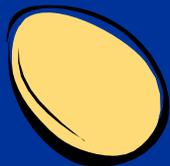
TGA on Plain Film

Companion Patient #2: Classic triad

Portable AP Plain Film, 1 day old patient

Classic triad:

1. Mild cardiomegaly
2. Mildly increased pulmonary vascular markings
3. “Egg-on-side” appearance to cardiac silhouette

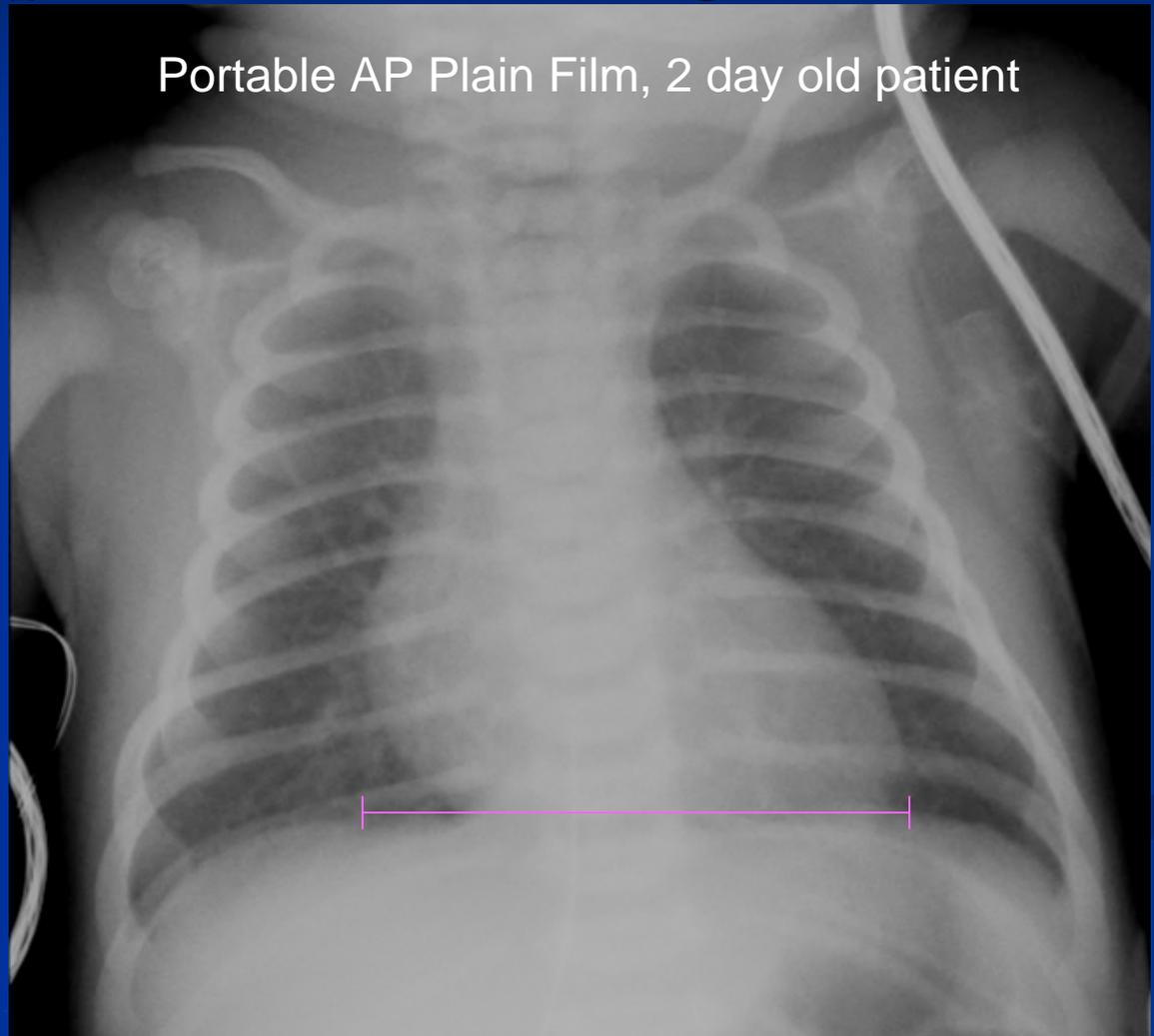


TGA on Plain Film

Companion Patient #3: Normal pulmonary markings

1. Mild cardiomegaly
2. Normal pulmonary vascular markings

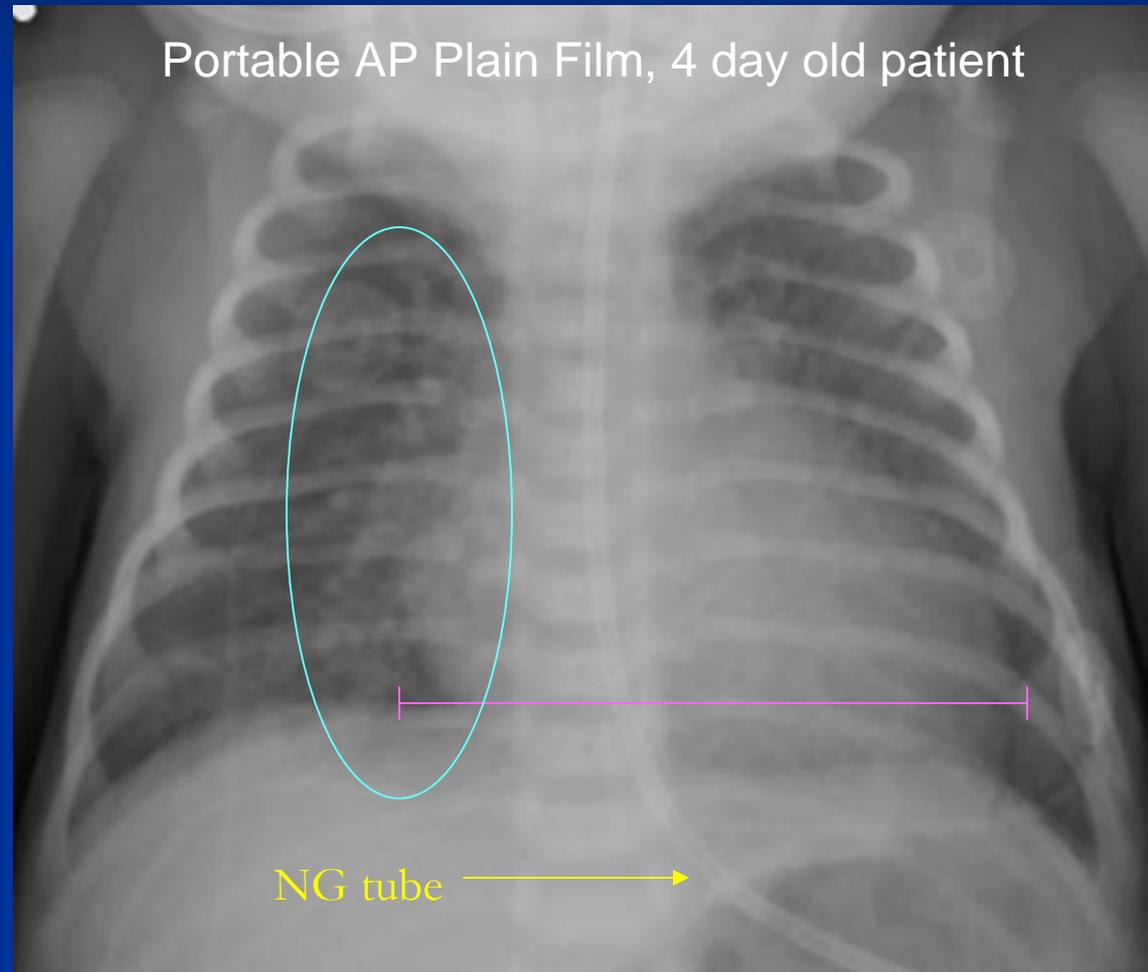
Note: the CXR can look completely normal in TGA!



TGA on Plain Film

Companion Patient #4: Prominent pulmonary markings

1. Mild cardiomegaly
2. Prominent pulmonary vasculature
3. NG tube



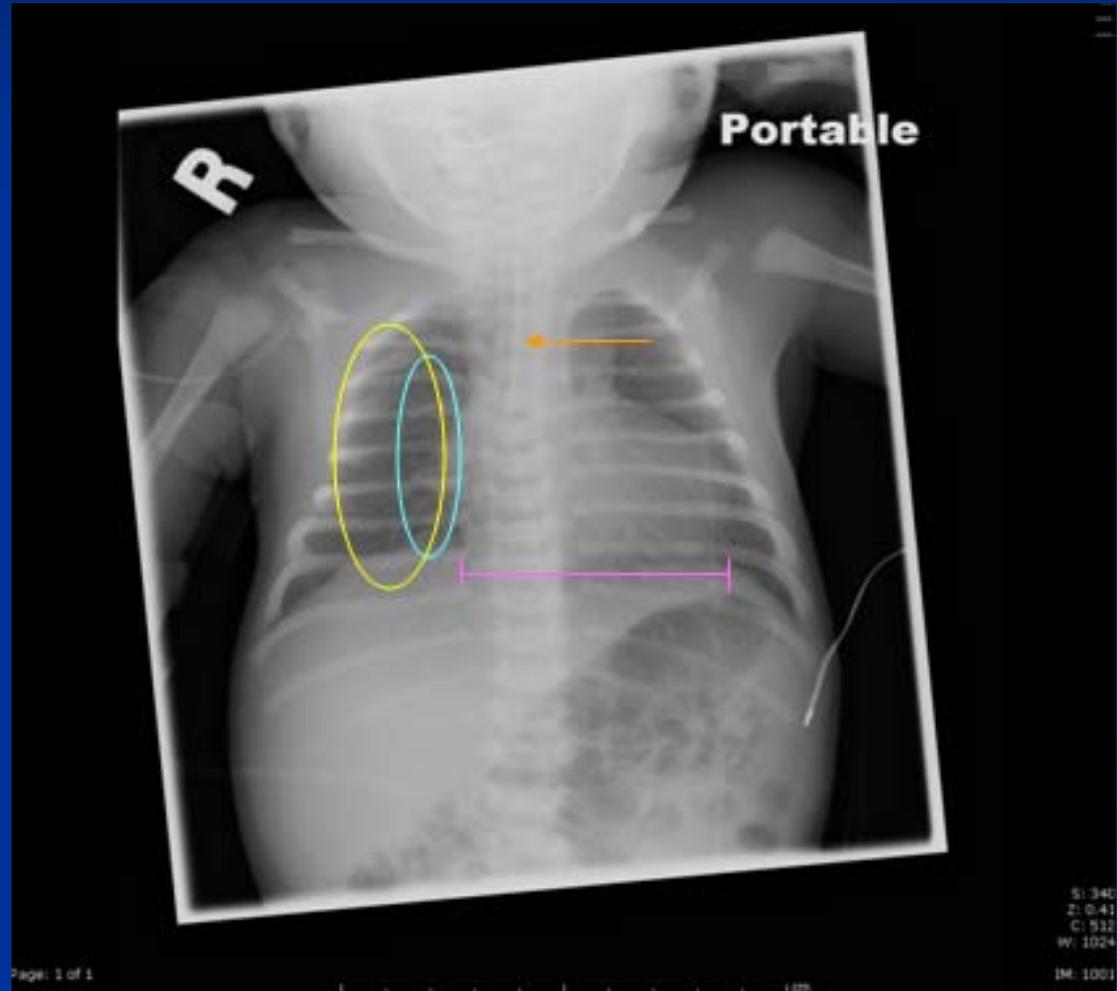
Baby L's CXR

Diagnosis: Meconium Aspiration

Portable AP Plain Film, 1 day old patient

Findings:

- Central pulmonary vasculature slightly prominent
- Very mild, diffuse groundglass opacification bilaterally
- Heart size within normal limits
- ET tube hovering over right mainstem bronchus (needs repositioning)



Dx: Meconium aspiration

Re-thinking Baby L's Diagnosis

Meconium aspiration syndrome:

- More common in post-term infants
- Clinical: cyanosis, tachypnea usually soon after birth
- CXR: streaky, linear densities, often centrally located; may see diffuse patchy densities

However, Baby L continued to have low O₂ saturation despite ventilation

- Poor response to supplemental oxygen suggests: shunt.
- A *cardiac* problem, not a primary *respiratory* problem, was now suspected, so an echocardiogram was performed...

Differential Diagnosis of Neonatal Respiratory Distress: Expanded

1. Aspiration of meconium or amniotic fluid
2. Congenital heart disease, especially cyanotic
3. Diaphragmatic hernia
4. Hyaline membrane disease/bronchopulmonary dysplasia
5. Pneumonia
6. Pulmonary immaturity
7. Respirator therapy (PEEP)
8. Transient tachypnea of the newborn

- Tetralogy of Fallot
- Complete transposition of the great arteries
- Truncus arteriosus
- Hypoplastic right or left heart syndrome
- Pulmonary atresia
- Persistent fetal circulation
- Asplenia syndrome (aka Ivemark syndrome)
- Ebstein anomaly

Echocardiography: Technique

- Echo = ultrasound of the heart.

Technique:

- Transducer is placed in a variety of locations
- For pediatric cardiology purposes, the “footprint” of the transducer is smaller to get images from between the ribs
- Study typically done by cardiologists

Echocardiography: Advantages and Limitations

Advantages:

- Non-invasive, safe, fast, relatively inexpensive, easily accessible (good for kids!)
- No radiation exposure
- Usually used to diagnose TGA – evaluates structure and function of heart, assess for other defects (VSD, LVOT obstruction)
- Use Doppler to help identify intracardiac and ductal shunts, valvular problems

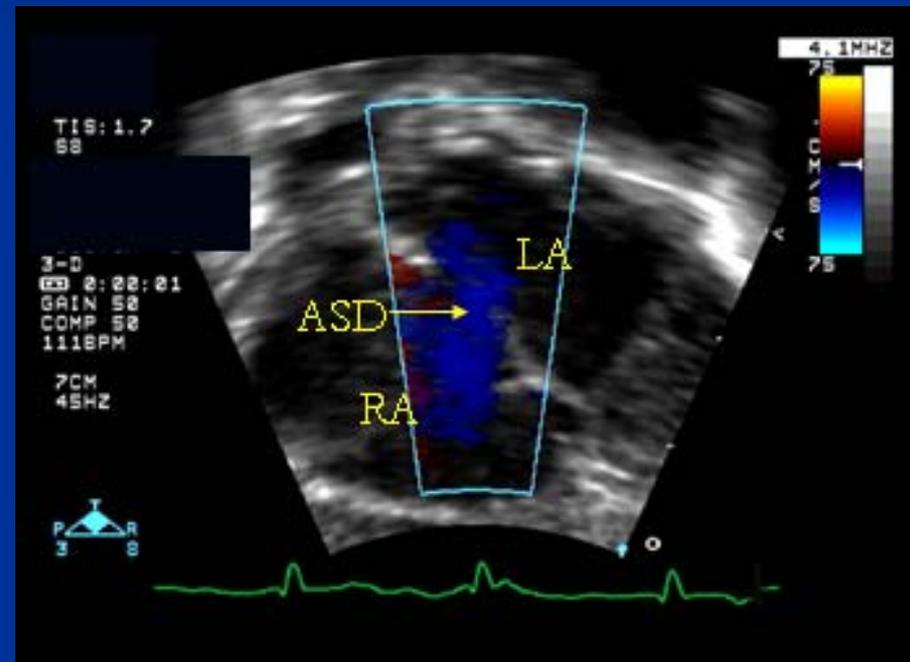
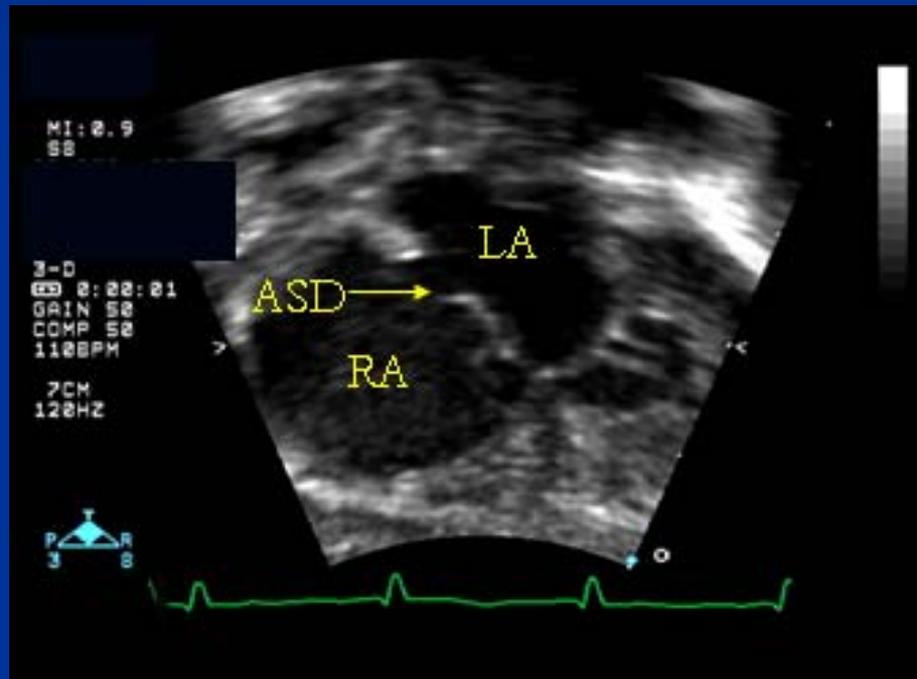
Limitations:

- Narrow field of view
- Operator dependent

TGA and Septal Defects on Echo: Introduction

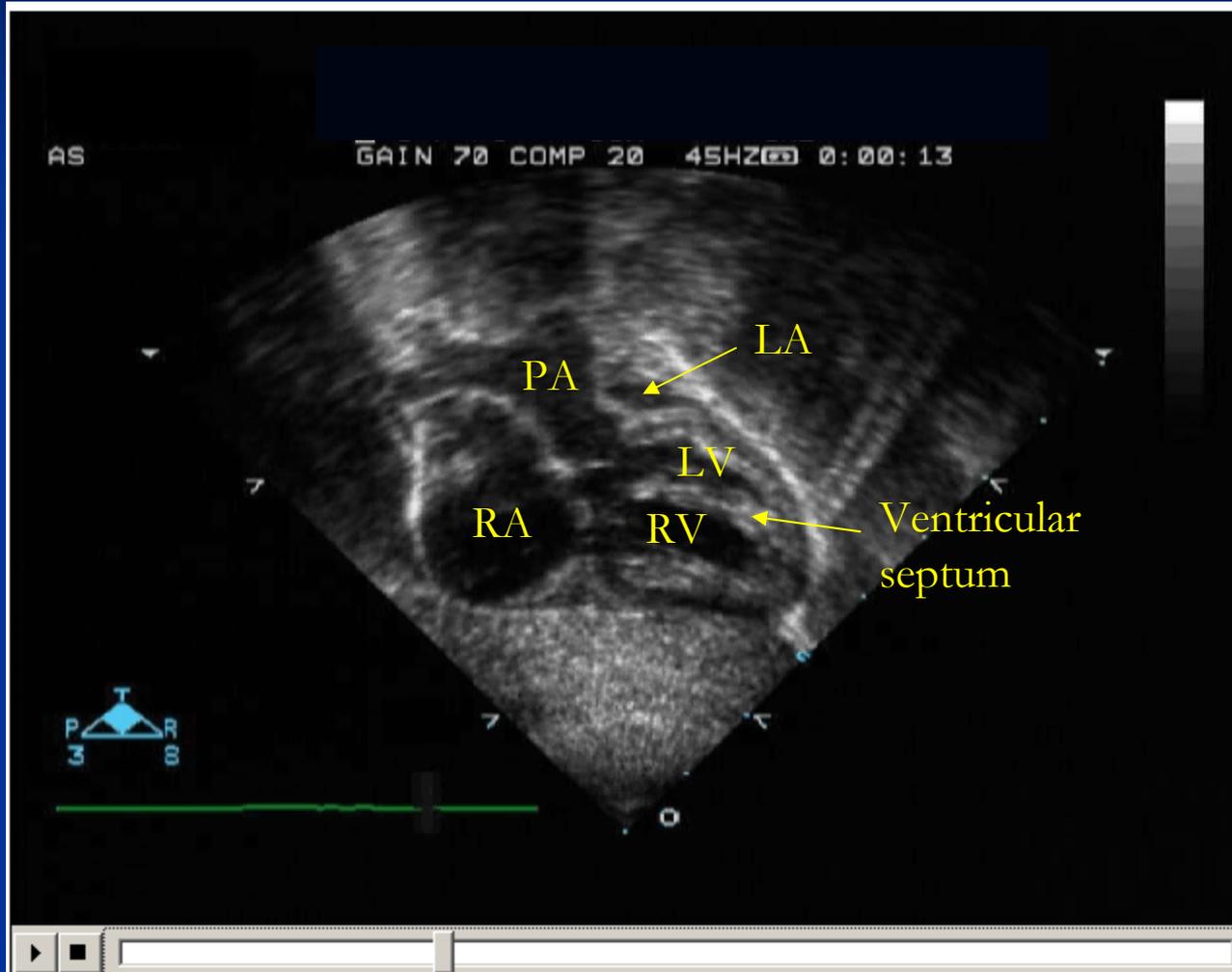
- Once again, we are going to view different patients with TGA on echo before viewing our patient's findings.
- We will look at a septal defect and classic TGA on echo, then look at Baby L's echo.

Companion Patient #4: Echo of ASD



Courtesy Dr. Barry Keane, Children's Hospital

Companion Patient #5: Echo of TGA



Courtesy Dr. Andrew Powell, Children's Hospital

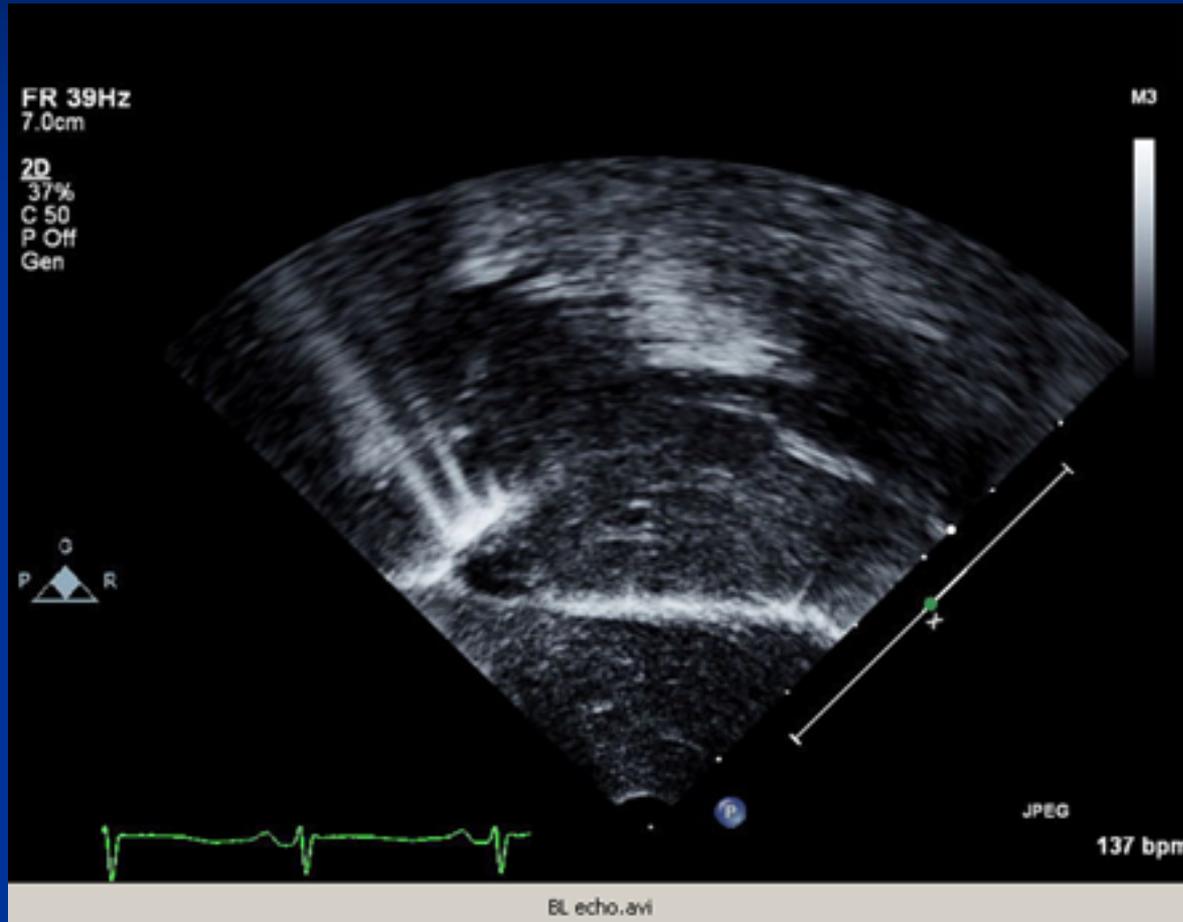
Companion Patient #5: Echo of TGA (Video)

- Posterior great artery arises from LV and divides into right and left pulmonary arteries
- Aorta is anterior and arises from RV



Baby L's Echo (Video)

Diagnosis: TGA



Courtesy Dr. Andrew Powell, Children's Hospital

Baby L's diagnosis changed to **Transposition of the Great Arteries**

The Next Step for Baby L: Cardiac Catheterization

- To perform balloon atrial septostomy
- To further characterize the defect in preparation for corrective surgery

Cardiac Catheterization and Cineangiography: Technique

Technique:

- Catheter inserted and guided to vessel in question, iodinated contrast injected, x-ray images obtained with a movie camera
- Terms: Arteriograms = images of arterial structures; venograms = images of venous structures; ventriculograms = images obtained after injecting contrast into a ventricle; coronary arteriograms = images obtained after injecting contrast into the coronaries

Cardiac Catheterization and Cineangiography: Advantages and Limitations

Advantages:

- Performed quickly in neonate for detailed hemodynamic information: establish diagnosis by angiography, identify major coexistent conditions, perform balloon atrial septostomy, evaluate post-operative complications
- Catheter course is diagnostic for TGA – can pass catheter from RA into aorta

Limitations:

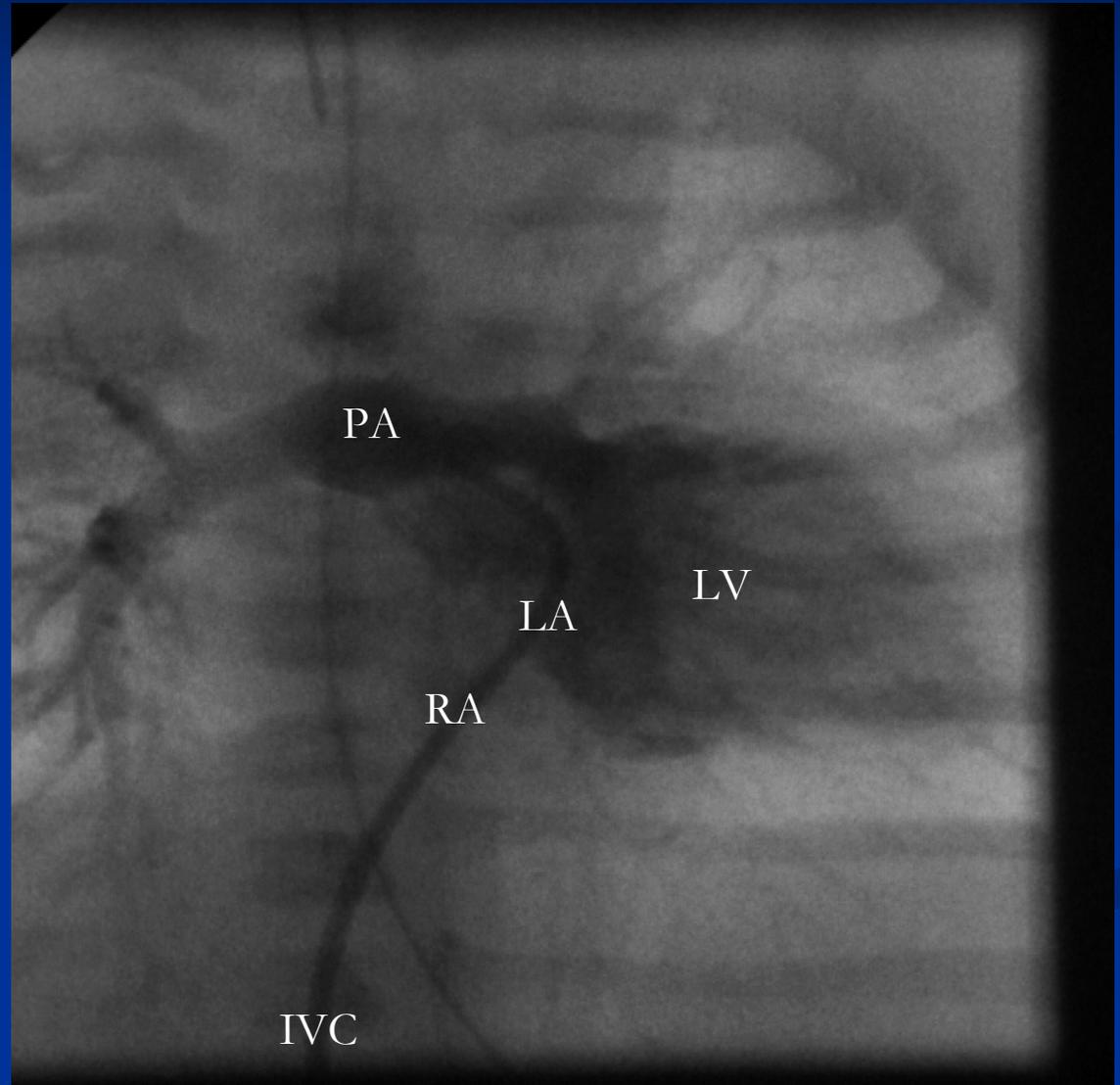
- Invasive
- Requires sedation

Companion Patient #6: Cineangiogram of TGA, AP View

Pulmonary arteriogram

AP View

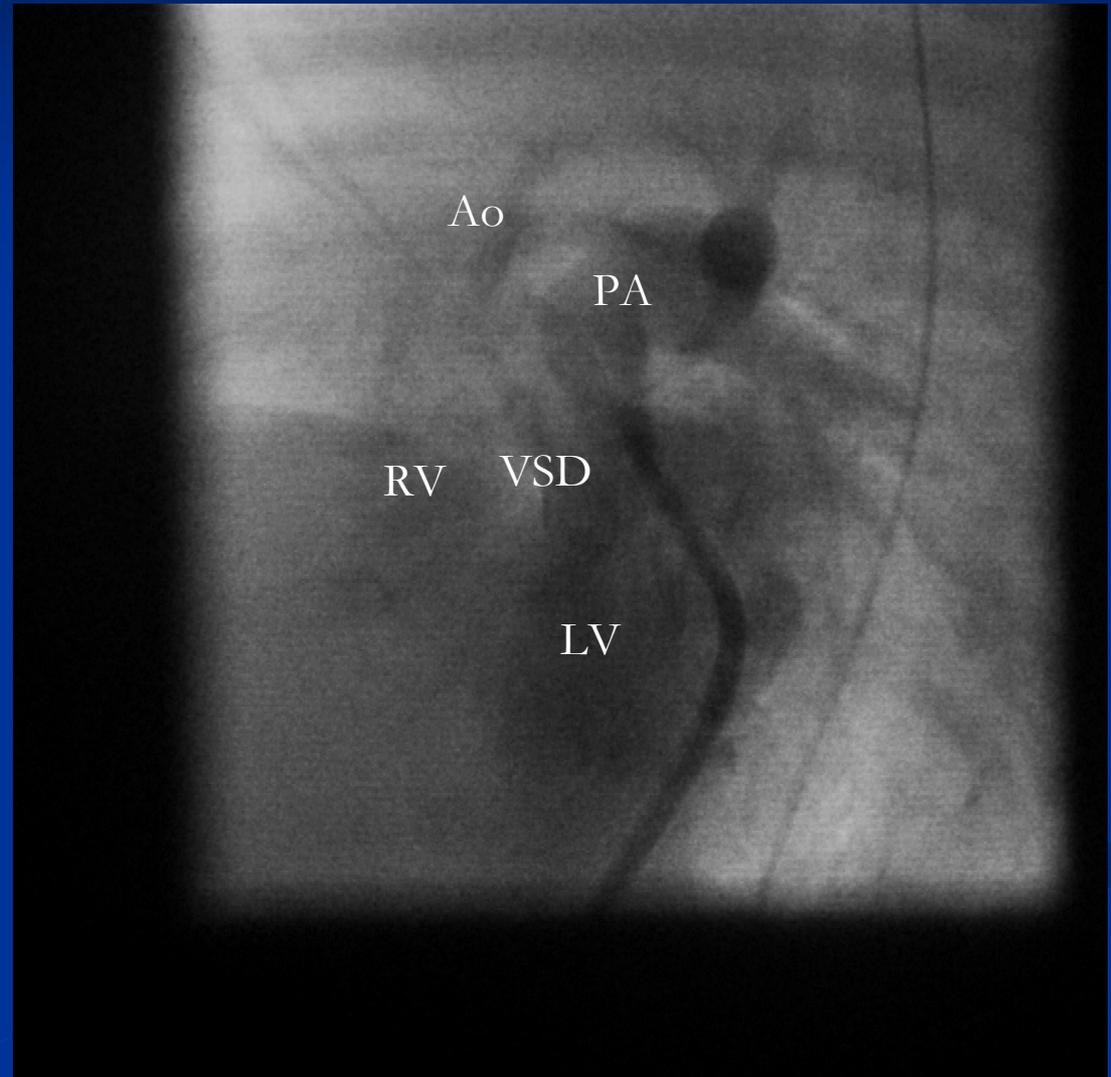
- Finding:
pulmonary artery
arising from LV



Companion Patient #6: Cineangiogram of TGA, Lateral View

Left ventriculogram Lateral-Oblique View

- Findings:
 - PA arising from LV, aorta arising from RV
 - VSD

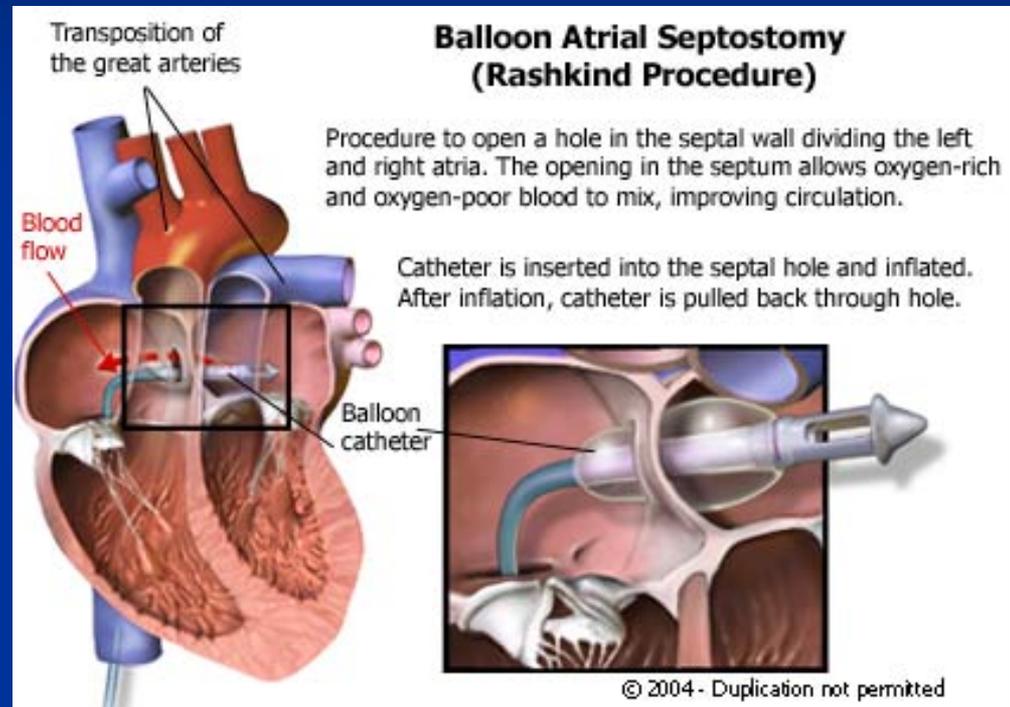


Treatment Options for TGA

- Balloon atrial septostomy
- Atrial switch operation
- Arterial switch operation
 - Complication: pulmonary stenosis

Balloon Atrial Septostomy (The Rashkind procedure)

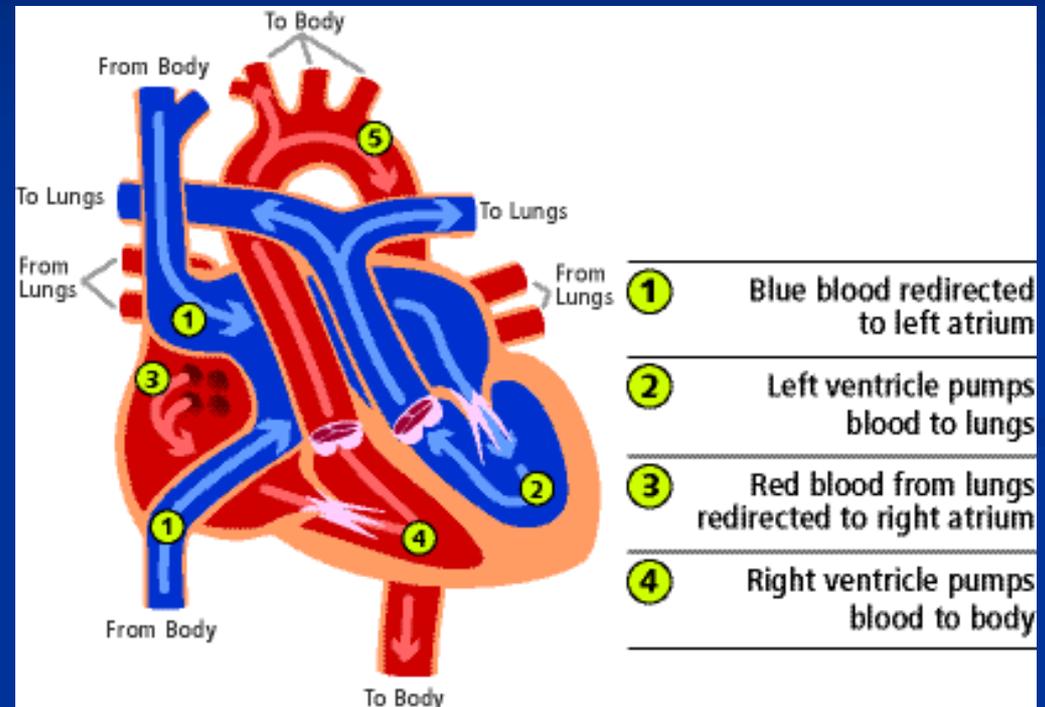
- Goal: increase interatrial communication to allow more mixing of the two circulations
- Increases circulation of oxygenated blood until heart defects can be surgically repaired
- Note: earliest treatment: prostaglandin E1 infusion to maintain PDA



<http://images.healthcentersonline.com/heart/images/article/Rashkind4.1.jpg>

Atrial Switch Operation (Mustard procedure)

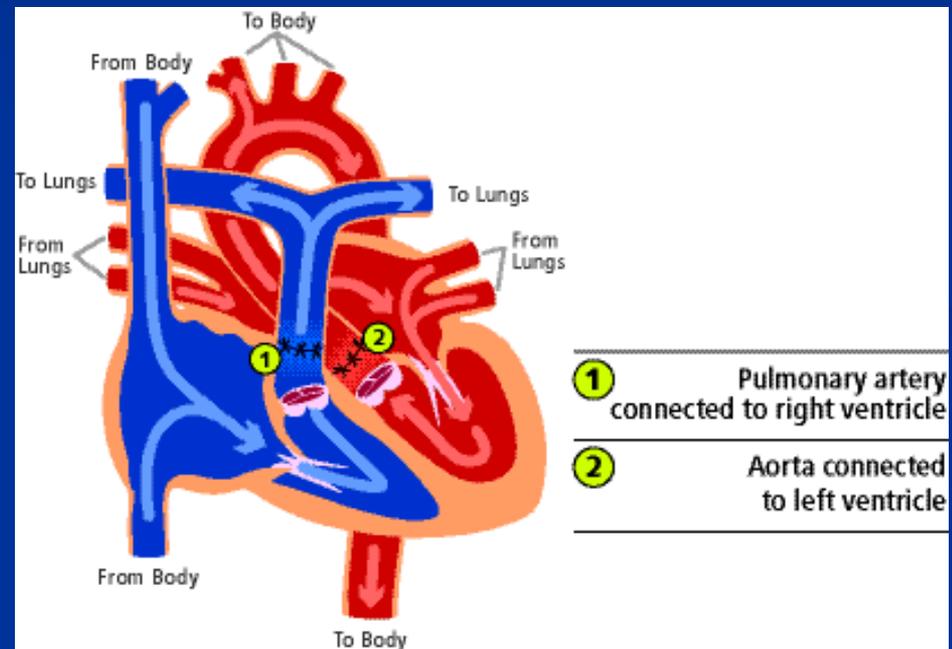
- No longer done
- Reroute venous return at the atrial level to the opposite ventricle, through creation of a “baffle” (=artificial obstruction to deflect flow)
- Problem: leaves RV and tricuspid valve in systemic circulation, so many patients later developed CHF due to RV dysfunction and/or tricuspid regurgitation
- Patients are still alive who have had this operation and are now presenting with sequelae of the operation



http://www.med.umich.edu/mott/chc/patient_con_tran1.html

Arterial Switch Operation (ASO) (Jatene procedure)

- First done in 1975, now preferred surgery to correct TGA
- Usually performed in first 2 weeks of life so LV does not get deconditioned from pumping into low resistance pulmonary vessels (may wait longer if large VSD present)
- Mortality from the operation is <5% (higher if VSD)
- Most common complication: supra-valvar pulmonary stenosis related to the reconstruction of the pulmonary artery



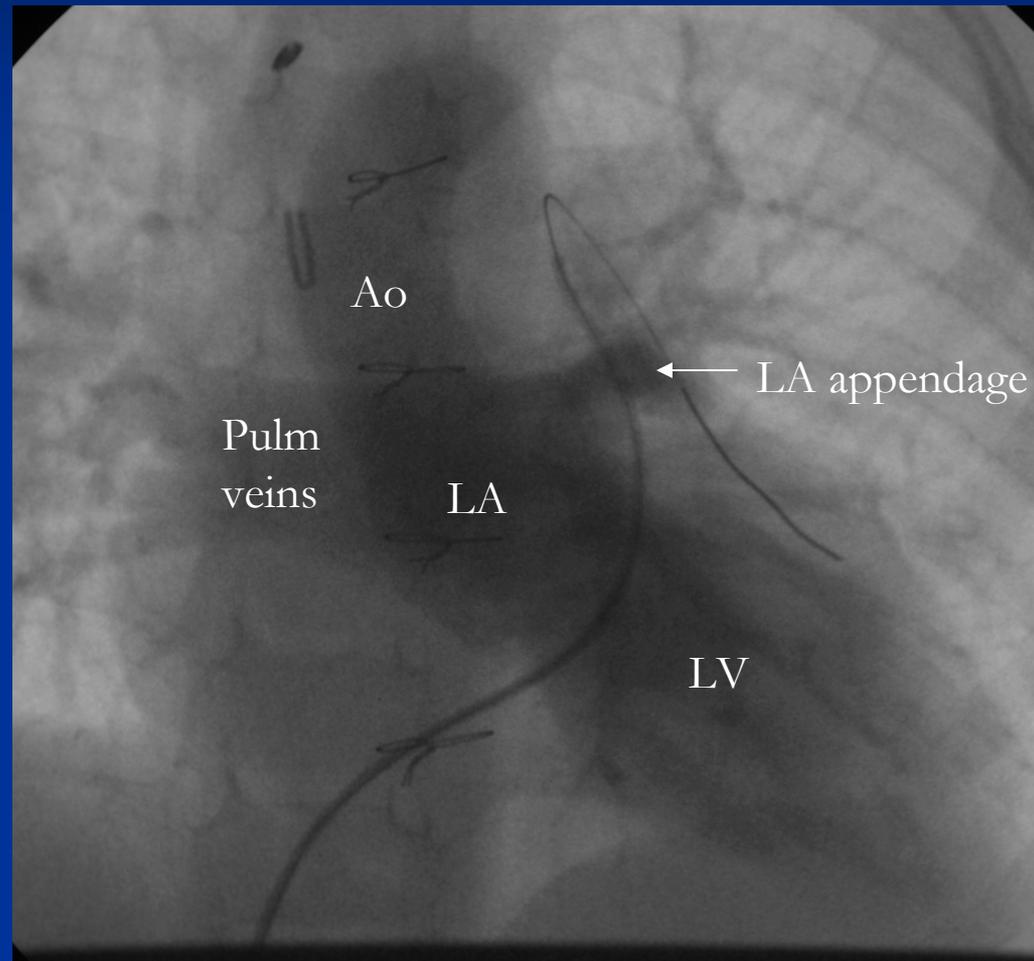
http://www.med.umich.edu/mott/chc/patient_con_tran1.html

Companion Patient #7: Post-ASO Cineangiography

AP View

Findings:

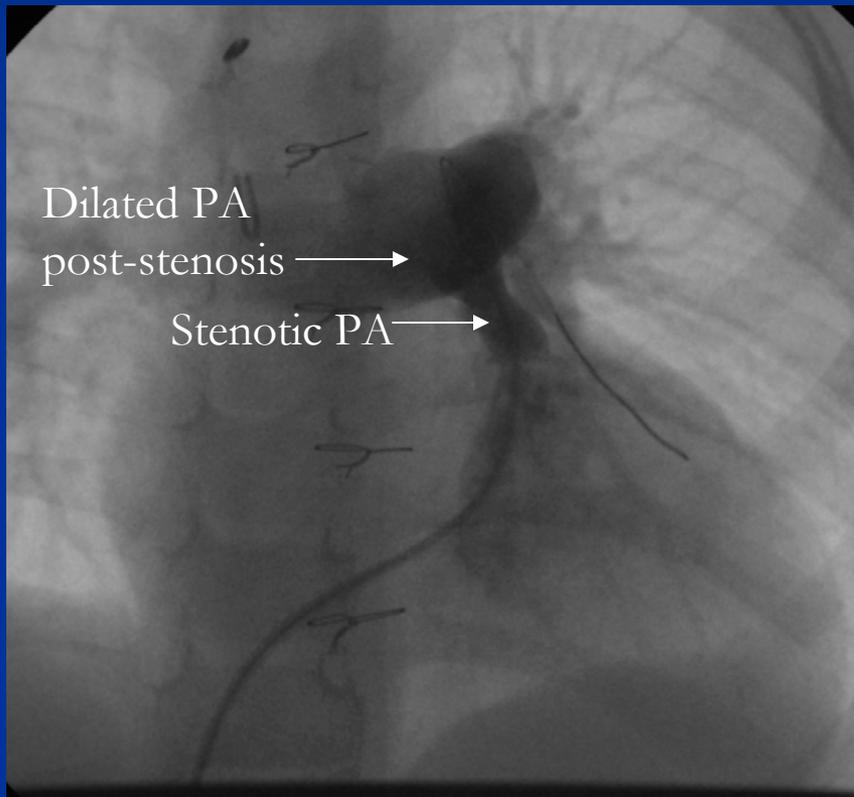
- Aorta arising from LV
- Note: After arterial switch operation, cineangiography looks fairly normal



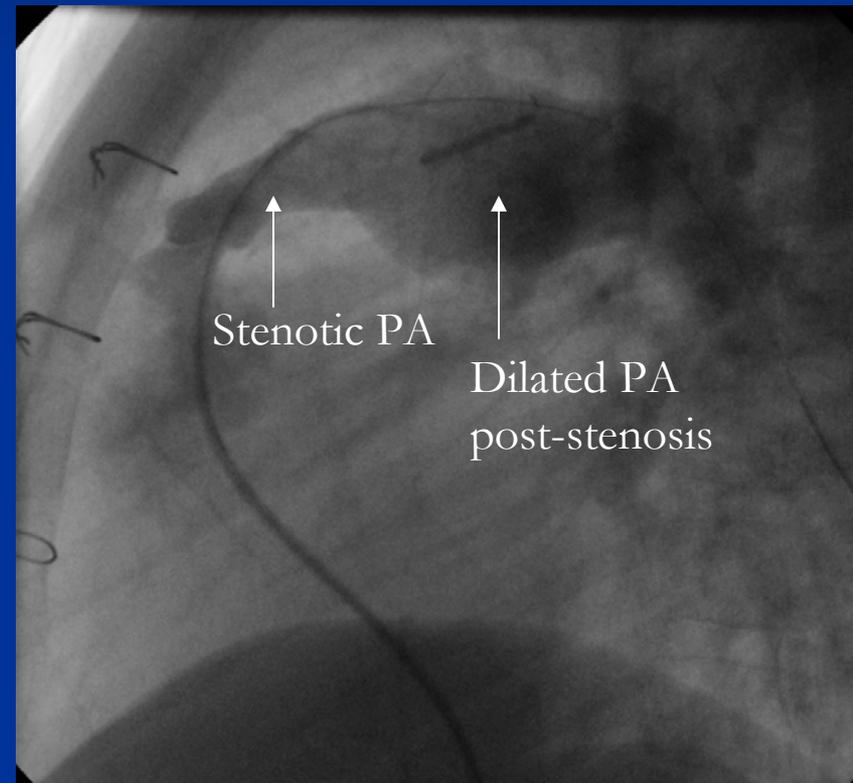
Courtesy Dr. Barry Keane, Children's Hospital

Complication of ASO: Supravalvular Pulmonary Stenosis Companion Patient #8: Cineangiogram

AP View



Lateral View

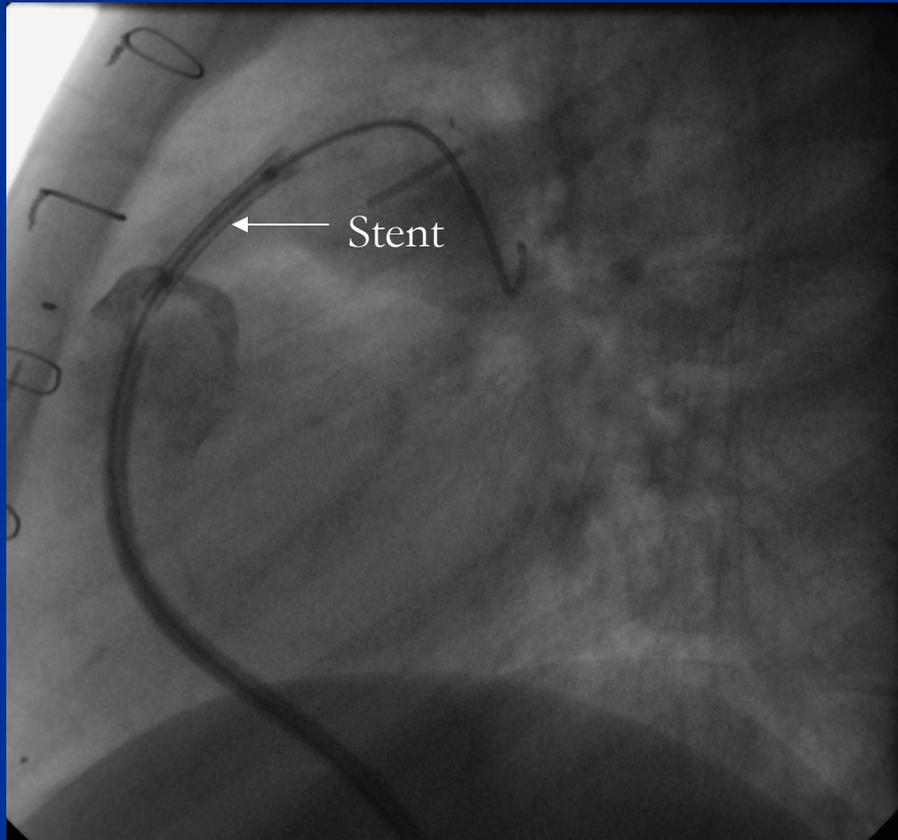


Courtesy Dr. Barry Keane, Children's Hospital

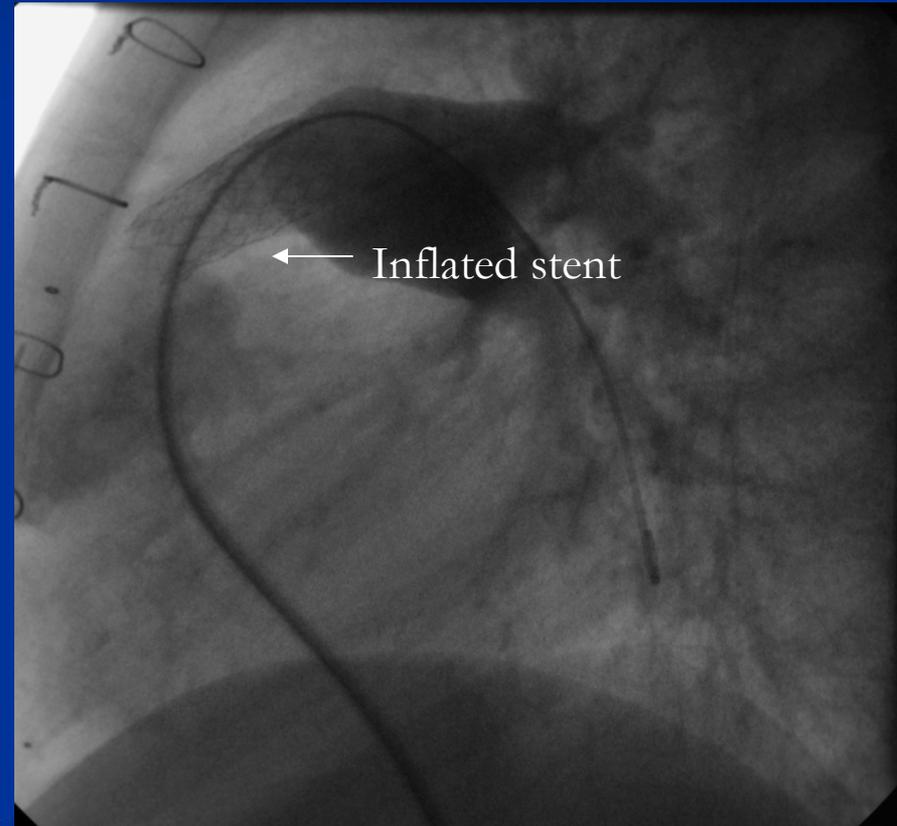
Treatment of Pulmonary Stenosis with Stent Placement

Companion Patient #8: Cineangiogram

Lateral View, Pre Inflation



Lateral View, Post Balloon Inflation



Courtesy Dr. Barry Keane, Children's Hospital

Follow-up on Baby L

- On Day 2 of life: After the diagnosis of TGA, Baby L received an echo-guided balloon atrial septostomy in the ICU.
- On Day 3: Baby L underwent an arterial switch operation.
- Currently, Baby L is 4 days old and is recovering from his ASO in the cardiac ICU.

Summary

- Think of transposition of the great arteries in a “blue baby”
- Diagnosis done prenatally (parallel great vessels) or by echo
- CXR can be deceiving!
- Cineangiography or echocardiography can be used for the balloon atrial septostomy
- Arterial switch can be lifesaving!

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