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Congenital Heart Disease II: The Repaired Adult

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Echocardiography Section, no disclosures
October 10, 2017; ASE Echo Florida

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Doc, I've had a heart operation….

Fontan

Rastelli

Jatene

Mustard

Amplatzer Closure

Senning

BTT shunt

Potts Shunt

Waterston shunt

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ACHD: Challenges in patients with repaired disease

- Repaired complex CHD is not “cured”
  - Patients require long term monitoring for sequelae of early surgeries
- Most important to understand the operative anatomy
  - Document procedural history
  - Review other imaging

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Case: 50 year old male history of tetralogy of Fallot, s/p repair age 7, CAD s/p stenting, aflutter ablation new patient evaluation

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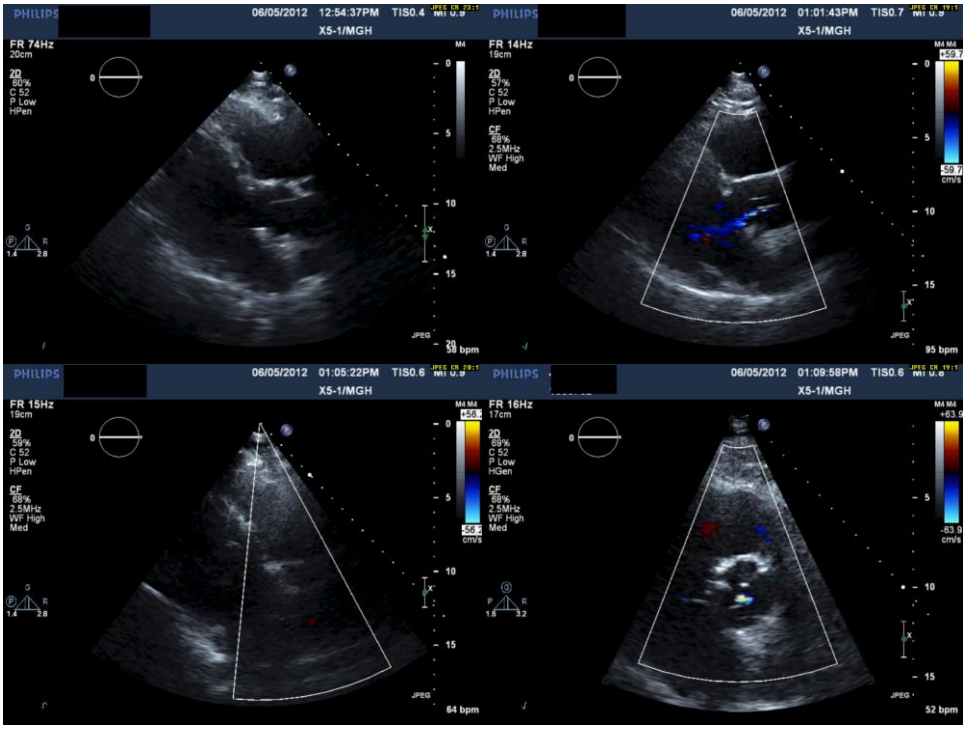
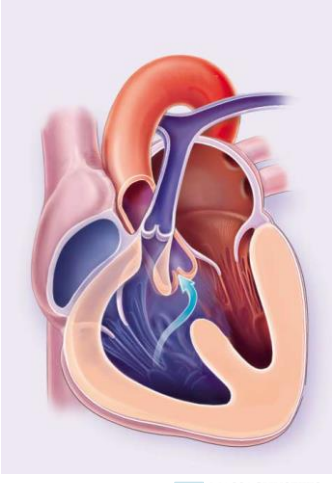


Tetralogy of Fallot: 4 features

- Rightward deviation ** of the aortic valve with overriding of the ventricular septum
- Ventricular Septal Defect
- Subpulmonary infundibular stenosis
- Right Ventricular Hypertrophy

- ** anterocephalad deviation of the outflow septum



Brickner NEJM 2000

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Associated abnormalities:

- Varying degrees of RVOT obstruction/PA hypoplasia
  - Mild obstruction: pink tetralogy
  - Most severe form: pulmonary atresia, cyanosis
- ASD (pentalogy)
- Right sided aortic arch (25%)
- Absent left pulmonary artery
- Coronary anomaly:
  - LAD from RCA, courses anteriorly across RVOT
  - Anomalous circumflex

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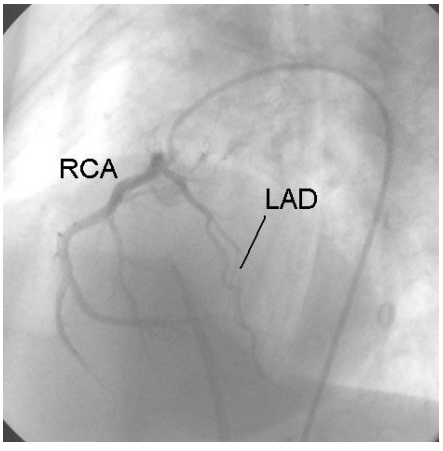

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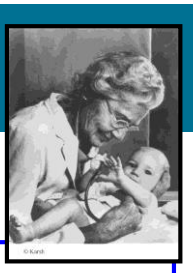
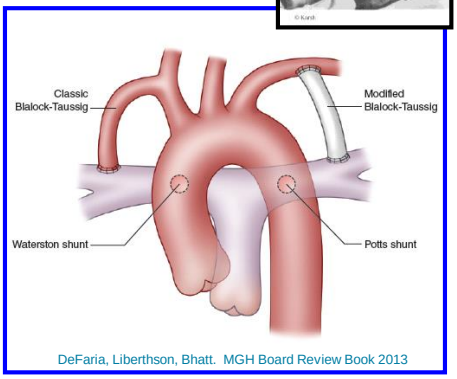
Palliative Systemic-Pulmonary Shunts



- **BTT Shunt:** Alfred Blalock, Helen Taussig, Vivian Thomas
  - 1944 : left subclavian artery to the left pulmonary artery
- **Potts:**
  - 1946 : descending thoracic aorta and LPA
- **Waterston:**
  - 1962 : ascending aorta to the RPA



DeFaria, Liberthson, Bhatt. MGH Board Review Book 2013

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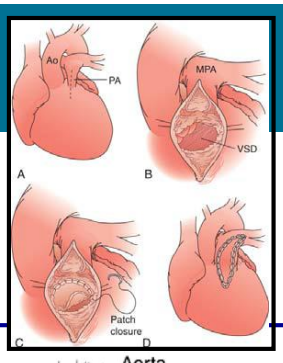
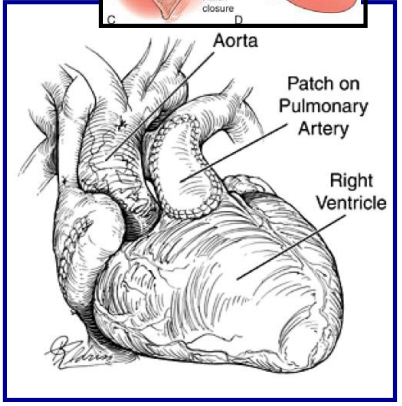
Definitive Surgical Correction:



- Complete Repair
  - 1954 C. Walton Lillehei (Hopkins)
  - 1955 John Kirklin (Mayo Clinic)
- Takedown of prior palliative shunt
- Open RVOT/PA
- VSD closure with patch
- Resection of subpulmonic obstruction
- Transannular patch developed in 1959
- Conduit between RV to PA in 1965 for pulmonary atresia



REPAIRED: NOT CURED
Sequelae post TOF repair

- Residual lesions (VSD, sub-PS, branch PA stenosis)
- **Pulmonary regurgitation**
- Progressive RV dilation and dysfunction: RHF
- Exercise intolerance/fatigue
- Ventricular arrhythmia and sudden death
- Secondary LV dysfunction
- Aortic dilation, insufficiency (dissection is quite rare)

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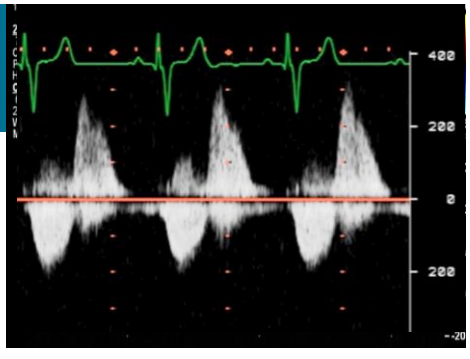
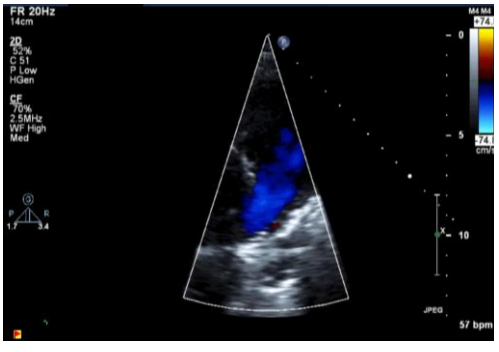
Pulmonic Regurgitation:



- **Often underappreciated**
- May be progressive over decades
- Volume load on the RV
- Color Doppler
  - Wide jet occupying >2/3 RVOT
  - Reversal of flow in the main PA
- CW Doppler:
  - Rapid deceleration time
  - Ends before end diastole



Indications for PVR in patients with repaired TOF

- RV size and function (cardiac MRI):
  - RVEDVI > 150ml/m2
  - RVESVI > 80ml/m2
  - RVEF < 47%
- LV systolic dysfunction: LVEF < 55%
- Large RVOT aneurysm
- QRS duration > 180ms
- Sustained tachyarrhythmias

Geva. *Circ* 2013

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Don't wait too long to fix PR….

- PVR done at RVEDVI of 195ml/m2….
- Severe residual RV dysfunction→ secondary LV dysfunction and cirrhosis
- Underwent Heart/Liver transplant

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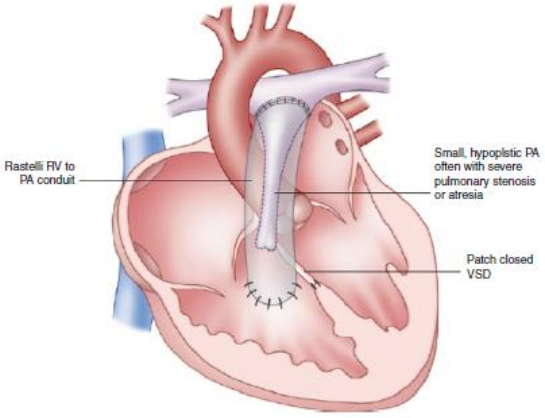
Small Pulmonary Arteries?
Rastelli conduit: RV to PA conduit



DeFaria, Liberthson, Bhatt. 2013

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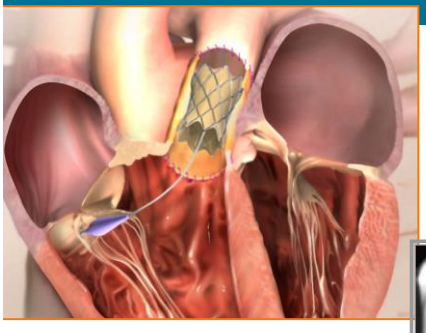
Transcatheter Pulmonary Valve



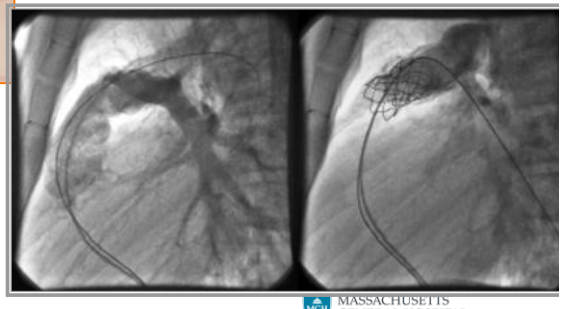


Image Courtesy of Inglessis

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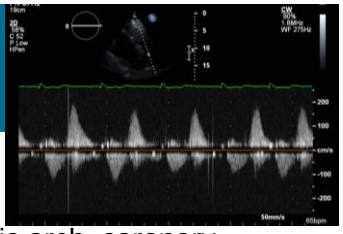
Summary: tetralogy of Fallot



- Think about associated anomalies (R aortic arch, coronary anomalies, ASD)
- Watch carefully for late sequalea (**PR**, RV dysfunction, VT)
- PVR timing: Don't wait too long….
  - RV function/size – Quantification is imperitive
- SCD risk assessment
- RV/PA conduits: percutaneous pulmonary placement may be considered

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D - Transposition of the Great Arteries:

32 M h/o D-TGA s/p Mustard atrial switch in infancy, presents with arial flutter

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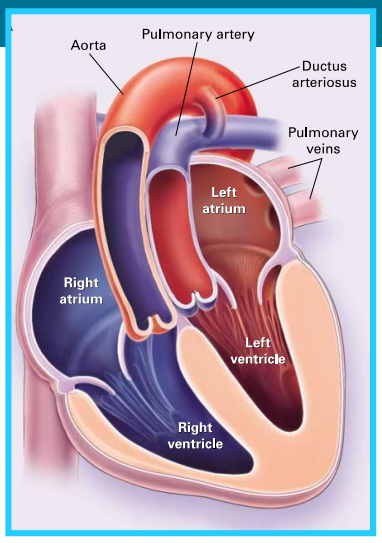
D-Transposition of the Great Arteries

- Normal ventricular situs
- Great arteries:
  - Failure of spiral septation of the truncus arteriosus
- Atrioventricular concordance
- Ventriculoarterial discordance
- 2 separate parallel circulations



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Brickner et al. *NEJM* 2000; 342: 334-342

Need for a shunt: atrial septostomy

VOLUME 90 JANUARY, 1950 NUMBER 1

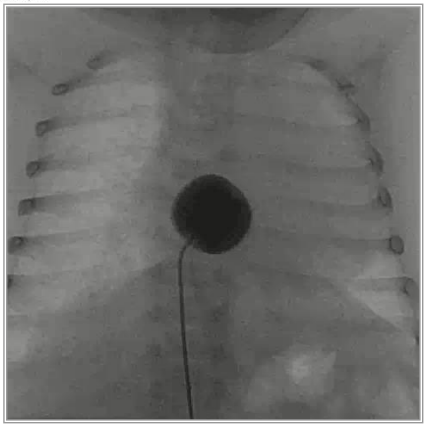
THE SURGICAL TREATMENT OF COMPLETE TRANSPOSITION OF THE AORTA AND THE PULMONARY ARTERY

ALFRED BLALOCK, M.D., F.A.C.S., and C. ROLLINS HANLON, M.D., Baltimore, Maryland

Creation of an Atrial Septal Defect Without Thoracotomy

A Palliative Approach to Complete Transposition of the Great Arteries

William J. Rashkind, MD, and William W. Miller, MD



1965

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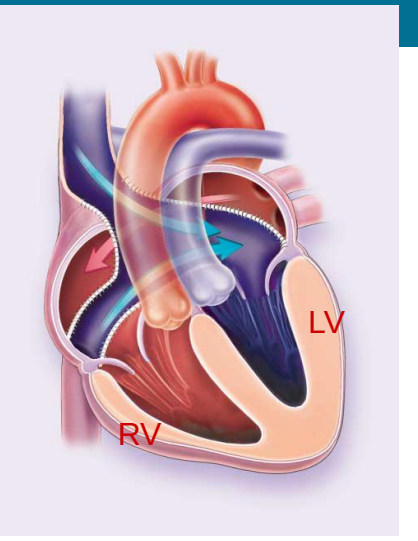
Atrial Switch Procedure

- 1958: Ake **Senning**
  - Atrial tissue
- 1964: William **Mustard**
  - Excised atrial septum
  - Synthetic material



Brickner, *NEJM* 2000

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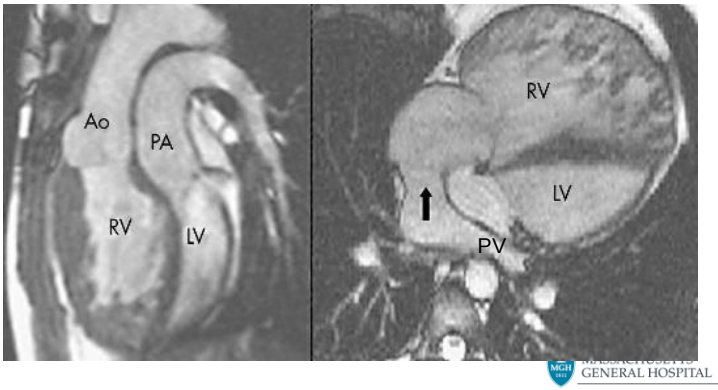
Atrial Switch: The Systemic Right Ventricle

- D TGA: The systemic RV is dilated and severely hypertrophied.
- Atrial switch pulmonary venous pathway: LLPV drains into RA through the baffle



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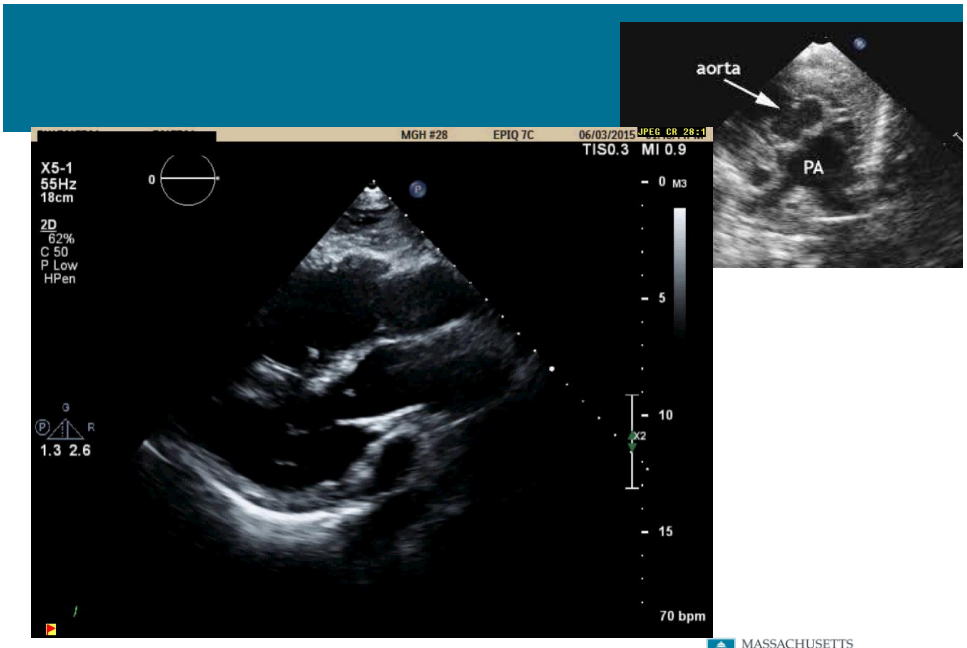

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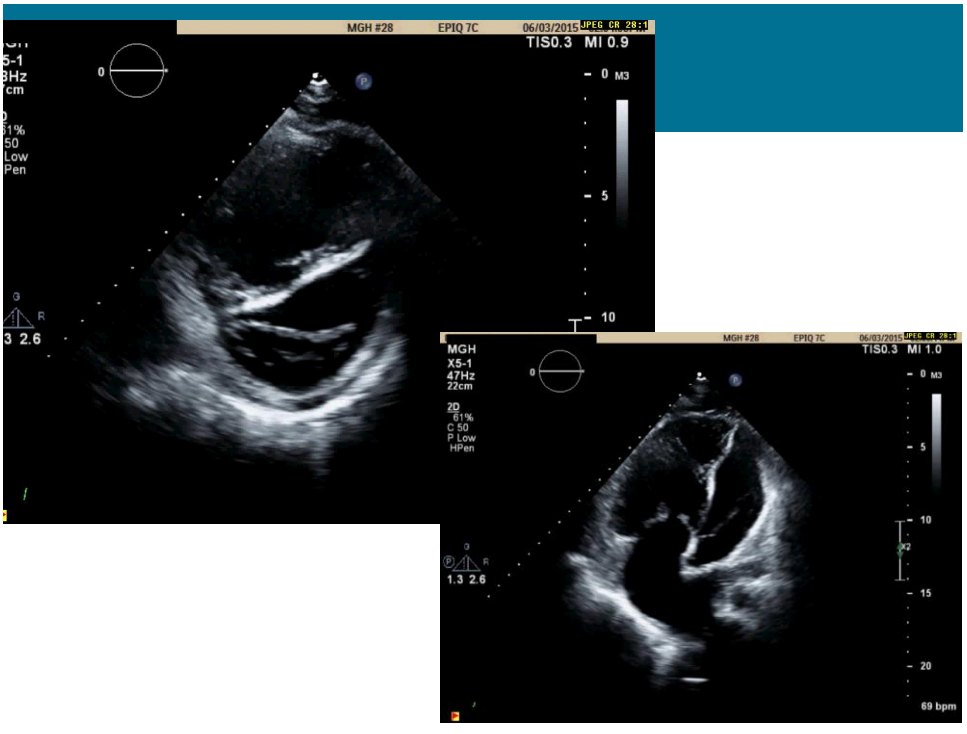

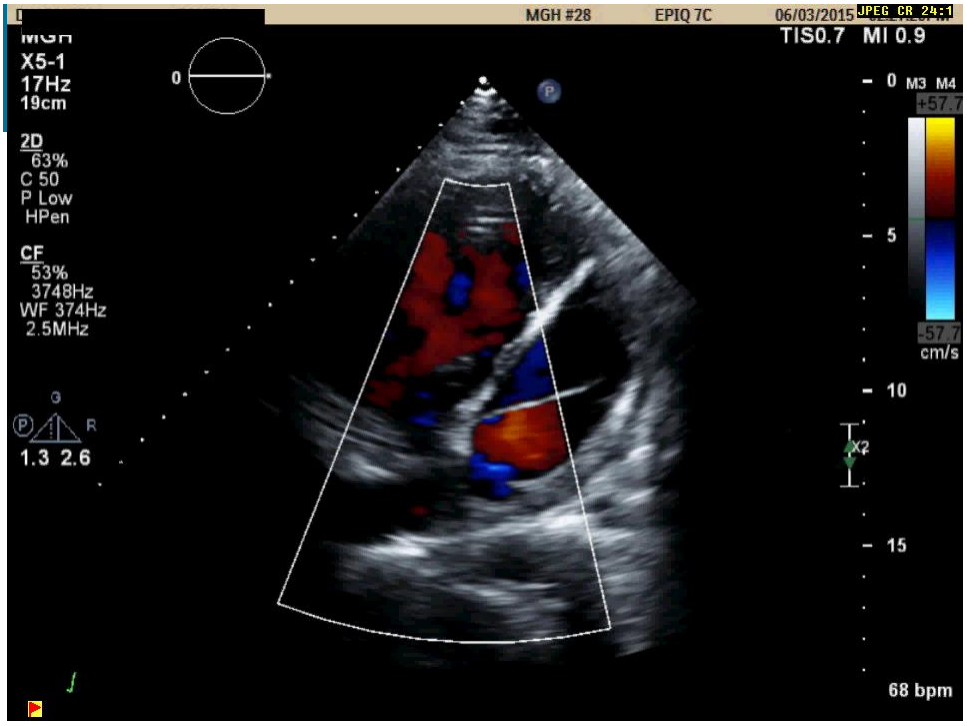

Issues after Atrial Switch

- **Arrhythmia**
  - Only 40% NSR @ 20 years post op, 11 % need pacers
  - Sudden death risk
    - Gelatt et al JACC 1997
- **Systemic RV failure**
  - RV systolic dysfunction -- ?ACE/ARB inhibitors - Hechter et al. AJC 2001
  - Progressive TR (systemic AV valve)
- **Venous baffle obstruction**
  - Mustard: SVC>IVC (SVC syndrome, hepatic congestion→ ascites)
  - Senning: Pulm vein>Systemic Veins
- **Baffle leaks (25%)**
  - Risk of paradoxical embolism

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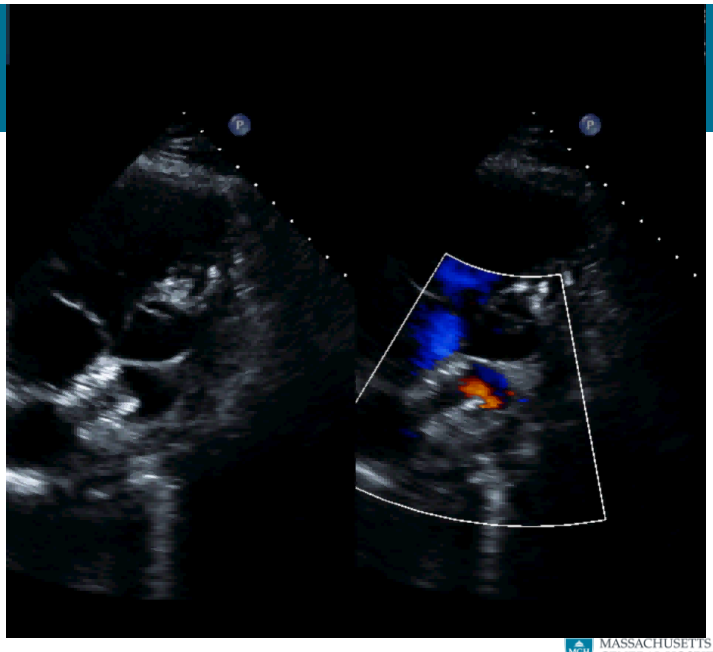
37M D-TGA s/p Mustard admitted with ascites, no orthopnea

- Lost to follow up for years
- Inferior< superior limb baffle obstruction
- Cirrhosis

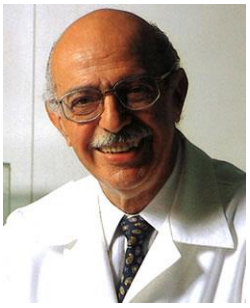
- Post baffle superior and inferior limb stenting
  - Autodiuresis
  - Improved ascites
- Regression of cirrhosis

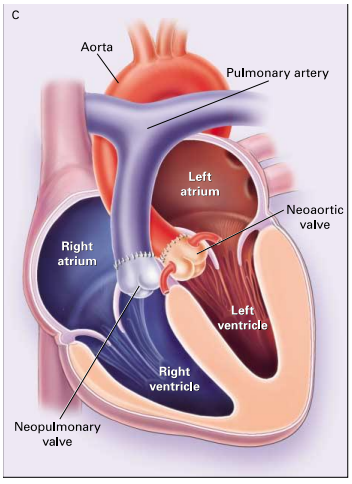


Arterial Switch Repair:
1975: Adib Domingos Jatene

Anatomic correction of transposition of the great vessels

Adib D. Jatene, M.D. (by invitation), V. F. Fontes, M.D. (by invitation), P. P. Paulista, M.D. (by invitation), L. C. B. Souza, M.D. (by invitation), F. Neger, M.D. (by invitation), M. Galantier, M.D. (by invitation), and J. E. M. R. Sousa, M.D. (by invitation), *São Paulo, Brazil*
Sponsored by E. J. Zerbini, M.D., *São Paulo, Brazil*





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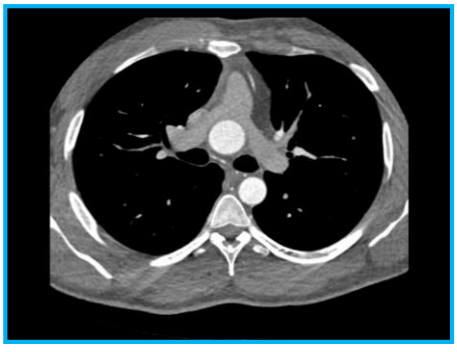
Brickner et al. NEJM 2000; 342: 334-342

Post-Op Jatene Arterial Switch



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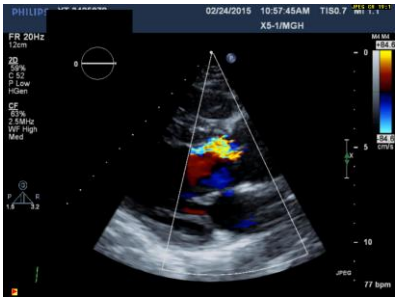
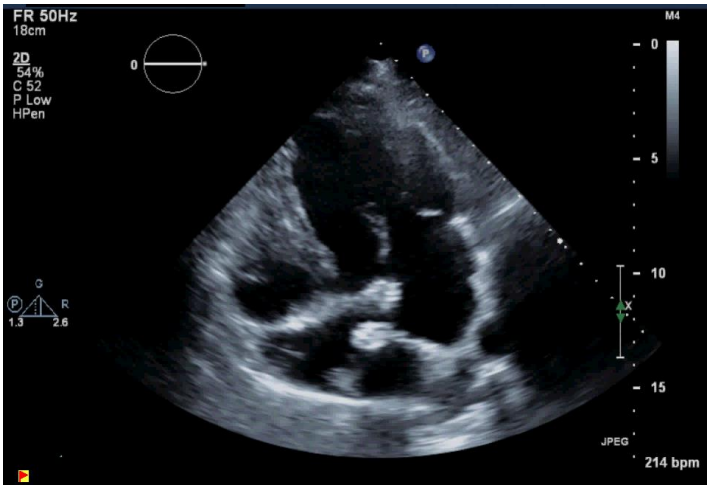
Problems after Arterial Switch Procedure

- Early:
  - Coronary insufficiency
  - PA stenosis after LeCompte
- Late:
  - Progressive aortic regurgitation
  - Neoaortic root dilation
- Generally:
  - Few long term rhythm or ventricular function issues



Pop Quiz…



Fontan Palliation: single ventricle repair

- Used in cyanotic heart disease
  - to separate blue from red
  - when a two ventricle repair is not possible
  - **and** when the PVR is low….

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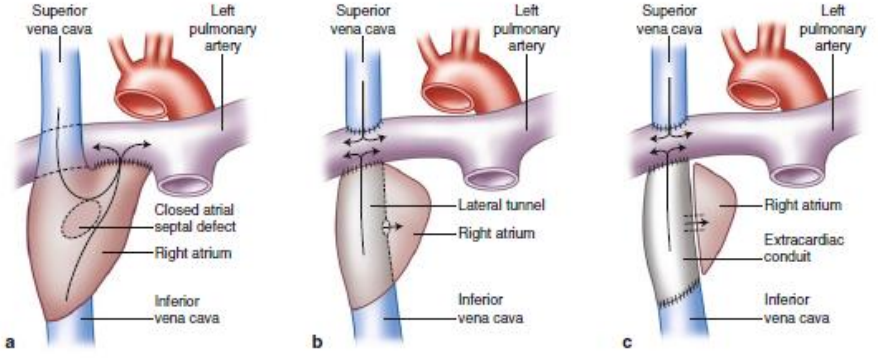
Fontan Palliation: Single Ventricular Physiology



Adapted from de Leval. *Nat. Rev. Cardiol. 2010*

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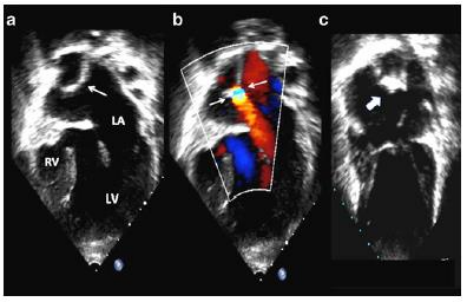
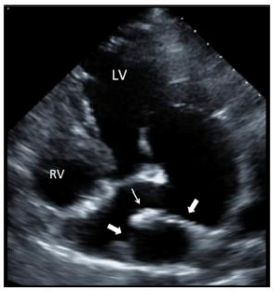
Tricuspid Atresia: status post lateral tunnel Fontan





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Summary:

Potts Fontan Rastelli

- Be clear about procedural history– look for scars
- Actively think about screening for late sequelae and procedural complications

Jatene

- Don't hesitate to call for help!

Senning Mustard BT shunt

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Thank You

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