Doc, I’ve had a heart operation…. 

- Mustard
- Senning
- Waterston shunt
- Rastelli
- Jatene
- Amplatzer Closure
- BTT shunt
- Potts Shunt
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

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

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**
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)

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

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
Small Pulmonary Arteries?

**Rastelli conduit:** RV to PA conduit

Transcatheter Pulmonary Valve
Summary: tetralogy of Fallot

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

D - Transposition of the Great Arteries:

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

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

Need for a shunt: atrial septostomy

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

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

Creation of an Atrial Septal Defect Without Thoracotomy

A Palliative Approach to Complete Transposition of the Great Arteries

William J. Rudolph, MD, and William H. Miller, MD

1965
**Atrial Switch Procedure**

- 1958: Ake Senning
  - Atrial tissue

- 1964: William Mustard
  - Excised atrial septum
  - Synthetic material

Brickner, *NEJM* 2000

**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
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
  - Progressive TR (systemic AV valve)
    - Hechter et al. AJC 2001

- **Venous baffle obstruction**
  - Mustard: SVC>IVC (SVC syndrome, hepatic congestion → ascites)
  - Senning: Pulm vein>Systemic Veins

- **Baffle leaks (25%)**
  - Risk of paradoxical embolism
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. (inventor), T. P. Furlow, M.D., C. I. Assisi, M.D., P. P. Paglia, M.D.
By the intervention of S. C. S. Pousada, E. P. Vagner, M.D., M. S. D. Furlow, M.D.
and J. L. M. Jatene, M.D. (inventor).
Sponsored by F. J. Zaidan, M.D., Elu Pascal, Brazil

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

Post-Op Jatene Arterial Switch
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....

Fontan Palliation: Single Ventricular Physiology

Adapted from de Leval. Nat. Rev. Cardiol. 2010
Tricuspid Atresia: status post lateral tunnel Fontan

Summary:

- Be clear about procedural history—look for scars

- Actively think about screening for late sequelae and procedural complications

- Don’t hesitate to call for help!
Thank You
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