Tennessee Chapter of ACC
Adult Congenital Heart Disease: Complex Thoughts on Simple Lesions & Simple Thoughts on Complex Lesions

Benjamin Frischhertz, M.D.
Assistant Professor of Medicine and Pediatrics
Director of the Adult Congenital Heart Disease Program
Vanderbilt Heart and Vascular Institute & Pediatric Heart Institute
11/3/18
CHD is Like Ice Cream
Managing CHD Involves Games of Strategy
Topics

• Holes (ASDs)

• Leaky pulmonary valves (repaired tetralogy of Fallot)

• Single ventricles (Fontans)
Holes (ASDs)
ASD types

- Superior sinus venosus defect
- Primum atrial septal defect
- Secundum atrial septal defect
- Inferior sinus venosus defect
- Coronary sinus defect
Large Ostium Secundum ASD
Secundum ASD Systematic Review

Closure of secundum ASDs in adults:

• Improved functional capacity
• Decreased RV size. RV systolic fxn unchanged
• Some evidence of increased LV size, increase LVEF ~ 5%
• Effect on mortality, incidence of AF unknown

Two important management questions for ASDs

• How can the defect be closed

• Should the defect be closed
How can it be closed
Device closure candidacy
Examples of ASD Closure Devices

- Amplatzer Septal Occluder
- Gore Cardioform
- Gore Helex
Should it be closed
ACHD Guidelines for ASD closure

- Symptoms attributable to ASD (Class I)
- Asymptomatic, but with significant right heart volume load – PASP < 50% systemic, PVR < 30% SVR (Class IIa)
- Net left-to-right shunt Qp:Qs > 1.5:1, PASP > 50% systemic, PVR > 30% SVR (IIb)
- No mention of paradoxical emboli in current ACHD guidelines
- **Contraindicated**: PASP > 2/3 systemic, PVR:SVR > 2/3, and/or net right-to-left shunt (III)

Stout et al. 2018 ACHD Guideline
Case

• 26y large secundum ASD, pulmonary hypertension, net left-to-right shunt
• Followed by ACHD, pulmonary hypertension
• On selexipag (prostacyclin receptor agonist) and tadalafil (PDE5 inhibitor)
Should the ASD be closed
Hemodynamics

Baseline:
Pressures:
RA 7 mmHg
PA 63/24, mean 37 mmHg
PCWP mean 8 mmHg
Ao 107/68, mean 81 mmHg

O2 Saturations:
SVC 69%
PA 84%
Aorta 98%
PV not sampled

Measured Hgb 13.2 g/dL
Assumed VO2 125 mL/min/m^2

Calculations:
Qp : Qs 2.1 : 1
Rp : Rs 0.19 : 1
PA pressure 58% systemic

On iNO:
Qp : Qs 3 : 1
Rp : Rs 0.13 : 1
PA pressure 57% systemic
Test occlusion

27 mm Amplatzer device
Post Closure

• D/c’d on antiplatelet, resumed pulmonary vasodilators

• Echo 3 mo later: PASP calculated 45 mmHg
Leaky Pulmonary Valves
(Repaired Tetralogy of Fallot)
Shaun White, tetralogy of Fallot
- 2 surgeries before age 1y
Long term outcome in repaired ToF

- 86% 32 year survival (96% in age/sex matched controls)
- 4% late sudden death (presumed to be VT)
- RV in TOF: 31% severe PI, 38% dilated RV (74% transannular patch repair)
- 20% with LV dysfunction (14% mild, 6% moderate or severe).

Murphy. NEJM. 1993.
Pulmonary Valves and Transannular Patch

Before

Post-op

PATCH
ACHD Guidelines for Pulmonary Valve Replacement

• PVR for repaired TOF and moderate or greater PI with symptoms (I)
• PVR in asymptomatic with moderate or greater PI and ventricular enlargement or dysfunction (IIa)
• PVR (and arrhythmia management) may be considered with moderate or greater PI and ventricular tachycardia (IIb)

Stout et al. 2018 ACHD Guideline.
Additional criteria for PVR

- Mild or greater RV or LV dysfunction
- Severe RV dilation (RV end-diastolic volume index ≥160 mL/m², RV end-systolic volume index ≥80 mL/m²)
- RV end-diastolic volume ≥2 times the LV end-diastolic volume
- RV systolic pressure two thirds or higher systemic pressure
- Progressive objective reduction in exercise capacity

Stout et al. 2018 ACHD Guideline.
TOF with pulmonary atresia

- 30y hx staged repair of TOF/PA including 16mm RV-to-PA homograft placement 1y
- s/p pulmonary valve replacement with 29mm Perimount 16y
- P/w DOE with minimal activity at age 30y
- Echo: prosthetic pulmonary valve with stenosis and insufficiency
RV 72/11, MPA 42/12, RPA 10/13, LPA 42/10

29 mm Edwards Sapien
Never had PVR, Repair “falling apart”  “Just right”  Too aggressive with PVR, PVR redo
Not Aggressive enough with PVR

• Hx TOF, transannular patch repair age 5y
• Presents to ACHD age 39y: dyspnea on exertion, atrial flutter
  – MRI: severe PI (RF 68%), RVEDVi 285 ml/m², RVESVi 185 ml/m², severe TR (RF 39%). LVEF 44%, RVEF 35%
  – EPS: required a-flutter ablation, VT inducible
  – Surgical PVR with 29mm Magna Ease Perimount
Cardiac MRI (age 39y)
Cardiac MRI (age 41y)

MRI age 42y: No PI, RVEDVi -124 ml/m², RVESVi 94 ml/m², mild TR, LVEF 64%, RVEF 25%
Cardiac MRI (age 48y)

Mild PI, Severe TR (RF 63%)

RVEDVi 278 ml/m^2, RVESVi 176 ml/m^2.

Tricuspid valve annulus 59mm

RVEF 37%
LVEF 50%

DOE, LE edema.
→ Medical management, permanent disability, ICD placed for primary prevention
Too aggressive?

51y repaired TOF:

- s/p BTT shunt (infant)
- s/p complete repair of tetralogy of Fallot (age 4y)
- s/p pulmonary valve replacement with Hancock procedure (age 9y)
- s/p PVR (age 11y)
- s/p redo PVR with St. Jude mechanical valve (age 17y)
- s/p redo PVR with 29mm Carpentier-Edwards bioprosthetic, tricuspid valve repair, PFO closure (age 39y)
- s/p dual chamber ICD placement
- history of atrial flutter s/p EP study with ablation

5 pulmonary valve replacements, avg. every 9y
Collateral Damage

- HCV +, requiring treatment
- Cirrhosis
- Variceal bleed
- Restrictive lung disease
- Scoliosis s/p instrumentation

Under consideration for heart and liver transplantation
Single Ventricles (Fontan)
Underlying lesions in Fontan

- Tricuspid atresia
- Hypoplastic left heart syndrome
- Double inlet left ventricle
- Pulmonary atresia intact ventricular septum
- Double outlet right ventricle
- Heterotaxy
- Abnormal tricuspid valve
- Atrioventricular canal defect
- Other

Anderson. JACC. 2008.
Types of Fontan

Modified classic Fontan

Lateral tunnel Fontan

Extracardiac Fontan

Fontan operation developed in 1971

Fontan – No physiologic right ventricle
“10 good years, 10 okay years, then trouble”
Extracardiac Sequelae of Fontan

33y history of heterotaxy, asplenia, unbalanced atrioventricular septal defect s/p single ventricle Fontan palliation.

- Previously unknown to ACHD
- Presented with hematemesis requiring transfusion
- Endoscopy showed varices, band acutely placed
Case

Alpha fetoprotein > 19,000
Fontan-Associated Liver Disease

- Chronic passive hepatic congestion
- Cirrhosis, ascites, synthetic dysfunction, portal hypertension, and hepatocellular carcinoma are possibilities
- Majority are asymptomatic (commonly: mild AST/ALT elevation, mild cholestasis pattern, mild INR elevation, thrombocytopenia)
- LFT’s not usually clinically helpful
- Biopsy?
- Effect on clinical management?

VanderbiltHeart.com
48y Fontan epicardial pacer
Fontan care takes an army, call for reinforcement
Vanderbilt ACHD Team
Questions / Comments

“To go boldly where no one has gone before”
Bibliography


Bibliography


Stout, Karen, et al. “2018 AHA/ACC Guideline for Adults with Congenital Heart Disease.” *JACC*. 2018