Adult Congenital Heart Disease

LAC+USC PGY2
December 4 and 11, 2019
ACHD

• Demographics and evolution of the specialty.
• Pregnancy and family planning.
• Repaired Tetralogy of Fallot (aka “Repaired Tet”).
• Functional Single Ventricle (aka “Single Ventricle” or “Fontan”).
Improved CHD Survival

Imaging and early diagnosis
Improved surgical and interventional techniques
Advances in critical care and EP

![Bar chart showing survival to 18 years of age with Moderate and Complex CHD for different years: 1940 (20%), 1960 (40%), 1970 (75%), 1980 (90%)].
Changing Proportion of Pediatric & Adult CHD

1965
- Pediatric patients, %: 30
- Adult patients, %: 70

1985
- Pediatric patients, %: 50
- Adult patients, %: 50

2005
- Pediatric patients, %: 60 (1 million)
- Adult patients, %: 40 (600,000)

Survival Curves for Tetralogy of Fallot stratified by decade of birth

![Graph showing survival curves for different decades of birth](image)
Morbidity and Mortality in ACHD

• After ~20 years from initial surgery, begin to see decreased survival, increased SCD, and increased hospitalizations.

• CHF and arrhythmia are biggest contributors to ACHD mortality.

Adult general cardiology fellowships require only 6 hours of ACHD education.

There are many ACHD patients with too few specialists and programs to take care of them.

>50% of CHD patients are lost to follow up after adolescence.

“Bethesda 32” 2000: ACC concludes the U.S. is not meeting the needs of the growing number of adults with CHD.
ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease): Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

AHA Scientific Statement

Best Practices in Managing Transition to Adulthood for Adolescents With Congenital Heart Disease: The Transition Process and Medical and Psychosocial Issues
A Scientific Statement From the American Heart Association

• 2011 Statement on Transition.
• Ensure continued follow up of CHD in adulthood.
• Assume responsibility for health care (independence).
• Exercise.
• Family Planning.
• Career Planning.
• Insurance.
ABIM ACHD Board Certification First Exams: 2015, 2017, 2019
Pediatric or Adult Cardiologists

ABIM OFFERS NEW ADULT CONGENITAL HEART DISEASE CERTIFICATE

Wednesday, July 30, 2014 - 11:15
The American Board of internal Medicine (ABIM) and the American Board of Pediatrics (ABP) are pleased to provide updated information on ABIM's new area of certification: Adult Congenital Heart Disease (ACHD), which is designed to recognize the qualifications of physicians who are specialists in the care of a wide range of adult patients with congenital heart disease.
22 ACHD Fellowship Programs in U.S.
2 years after pediatric or adult cardiology fellowship.
2015: ACHA Begins Formal Accreditation Process for Care Centers

ACHA Launches National Accreditation Program

Eleven Centers Throughout United States First to Earn ACHA Adult Congenital Heart Disease (ACHD) Accreditation

Individuals with congenital heart disease (CHD), the most common birth defect diagnosed in one in 100 births, are living longer. In fact, there are now 14 million adults in the U.S., more than children, currently living with one of the many different types of congenital heart defects that range among simple, moderate, and complex. To better serve this population, the Adult Congenital Heart Association (ACHA), the only organization in the country that specifically serves adults with CHD, announced the launch of the ACHA Adult Congenital Heart Disease (ACHD) accreditation program. This accreditation program will provide a community of support and network of experts with knowledge of the disease. To date, 11 centers have earned the ACHA ACHD Accredited Comprehensive Care Center designation:

- Ahmanson/UCLA Adult Congenital Heart Disease Center (Los Angeles, CA)
- Adult Congenital Heart Program, Stanford University (Palo Alto, CA)
- Colorado Adult and Teen Congenital Heart (CATCH) Program at Children’s Hospital Colorado and University of Colorado Hospital (Aurora, CO)
- Boston Adult Congenital Heart (BACH) and Pulmonary Hypertension Program (Boston, MA)
- University of Michigan Adult Congenital Heart Program (Ann Arbor, MI)
- Washington University Adult Congenital Heart Disease Program (St. Louis, MO)
- Adult Congenital Heart Disease Program at Children’s Hospital & Medical Center and Nebraska Medicine (Omaha, NE)
- Cincinnati Children’s Adult Congenital Heart Disease Program (CCHMC) (Cincinnati, OH)
- COACH: Columbus Ohio Adult Congenital Heart Disease & Pulmonary Hypertension Program (Columbus, Ohio)
- Adult Congenital Heart Disease Program at University of Washington & Seattle Children’s Hospital (Seattle, WA)
- Providence Adult and Teen Congenital Heart Program (PATCH) (Spokane, WA)
<table>
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<tr>
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<th>ACHA ACHD PROGRAM CRITERIA</th>
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<tbody>
<tr>
<td>A</td>
<td>ACHD Cardiologist</td>
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<td>B</td>
<td>ACHD Medical Program Director</td>
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<tr>
<td>C</td>
<td>Advanced Practice Nurse/Physician Assistant</td>
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<td>D</td>
<td>Registered Nurse</td>
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<td>E</td>
<td>Cardiothoracic Surgery and Cardiothoracic Intensive Care Unit</td>
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<tr>
<td>F</td>
<td>Heart Failure, Heart Transplant, Heart/Lung Transplantation</td>
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<td>G</td>
<td>Interventional Cardiac Catheterization</td>
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<td>H</td>
<td>Interventional Electrophysiology</td>
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<td>Patient-Centered Care</td>
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<td>Cardiac Magnetic Resonance Imaging</td>
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<td>Exercise Testing and Cardiac Rehabilitation</td>
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<td>Reproductive Services</td>
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<td>Psychology and Social Work</td>
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Cardio-facial Syndromes
Down Syndrome

- Trisomy 21
- ~40% have CHD.
- ~40% of CHD: Atrioventricular Septal Defect.
- Multi-organ system disorder.
Turner Syndrome

- Absent or abnormal X chromosome (45XO).
- Bicuspid aortic valve, coarctation, PAPVR, aortopathy.
- Short stature, premature ovarian failure.
Noonan Syndrome

- Turner-like phenotype.
- PTPN11 or DRAS gene mutation; autosomal dominant.
- Pulmonic stenosis, HCM, ASD.
22q11 Deletion Syndrome

• aka: DiGeorge, Velocardiofacial syndrome, Catch-22.
• Conotruncal abnormalities (TOF, TA, DORV).
• Autosomal Dominant.
• Multi-organ system involvement.
Genetic Syndromes
ACHD

• ~25% genetic or syndromic.
• Majority of isolated cases of CHD have no apparent cause.
Risk of CHD in Offspring

• General population: ~1%.
• First degree relative (of fetus) has CHD: ~4%.
• First degree relative (of fetus) with bicuspid aortic valve: ~9%.
• Mom or dad with autosomal dominant condition: 50% (Marfan, Williams, Holt-Oram, 22q, congenital long QT, HCM.)
Pregnancy: Increased C.O., Decreased SVR
Problem for those with:

• Poor functional capacity.
• Weak right or left ventricle.
• Cyanosis.
• Obstructive lesions, e.g. MS, HOCM, AS, severe RVOTO.
• Pulmonary hypertension.
Pregnancy: Hypercoagulable State
Problem for those with:

• Mechanical heart valve.
• Shunts (potential for paradoxical embolism).
• Functional single ventricle.
• Pulmonary hypertension.
Pregnancy Contra-indicated

- PAH.
- EF<30%.
- History of PPCMP with any residual LV dysfunction.
- Severe MS or severe symptomatic AS.
- Marfan >45 mm, BAV >50 mm.
- Severe native coarctation.
Table 1: CARPREG II Risk Predictors

<table>
<thead>
<tr>
<th>Predictor</th>
<th>Points</th>
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<tbody>
<tr>
<td>Prior cardiac events or arrhythmias</td>
<td>3</td>
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<tr>
<td>Baseline NYHA 3–4 or cyanosis</td>
<td>3</td>
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<tr>
<td>Mechanical valve</td>
<td>3</td>
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<tr>
<td>Systemic ventricular dysfunction LVEF&lt;55 %</td>
<td>2</td>
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<td>High-risk valve disease or left ventricular outflow tract obstruction (aortic valve area &lt;1.5 cm², subaortic gradient &gt;30, or moderate to severe mitral regurgitation, mitral stenosis &lt; 2.0 cm²)</td>
<td>2</td>
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<td>Pulmonary hypertension, RVSP &gt;49 mmHg</td>
<td>2</td>
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<tr>
<td>High-risk aortopathy</td>
<td>2</td>
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<td>Coronary artery disease</td>
<td>2</td>
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<tr>
<td>No prior cardiac intervention</td>
<td>1</td>
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<td>Late pregnancy assessment</td>
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Primary cardiac event risk: score = 1, 5 % risk; score = 2, 10 % risk; score = 3, 15 % risk; score = 4, 22 % risk and 41 % risk if score greater than 4. NYHA = New York Heart Association Functional Classification; LVEF = left ventricular ejection fraction; RVSP = right ventricular systolic pressure. Source: Silversides et al., 2018, with permission from Elsevier.¹⁴
Maternal Complications

Most Common

• Arrhythmia
• Heart Failure

Most Worrisome

• Thromboembolism
• Aortic dissection
Mode of Delivery

• Generally anticipate normal L & D.
• NSVD: Less bleeding, fewer clots, lower rate of infection vs C-section.
• C-section reserved for obstetric indications or:
  ➢ Therapeutic INR on warfarin (risk of fetal ICH with vaginal delivery).
  ➢ Aortic dissection.
  ➢ Enlarged aorta (MFS>40mm, BAV>45mm).
  ➢ Maternal instability.
  ➢ Before necessary surgery if >28 weeks.
Birth Control

• In some women with complex ACHD pregnancy may be life threatening (efficacy is critical).

• In some women with CHD estrogen containing birth control methods may increase the risk of life-threatening thrombosis (safety is critical).
Birth Control: Avoid Estrogen

• Estrogen increases risk of thrombosis by stimulating production of clotting factors.

• Absolutely contra-indicated: R to L shunt, mechanical valve, pulmonary arterial hypertension, EF <30%, Fontan circulation.

• Relatively contra-indicated: h/o thromboembolism, atrial arrhythmias, ASD.
Birth Control: Challenge

- 50% of women for whom pregnancy would be life threatening have not been advised to avoid pregnancy.

- Close to 50% of women with CHD for whom estrogen is contraindicated have used estrogen containing birth control methods.
TETRALOGY OF FALLOT
JB

• 26 year old man born with Tetralogy of Fallot
Tetralogy of Fallot: Anterior Displacement of the Conotruncal Septum

Tetralogy of Fallot

Major Defects
1. Pulmonary Stenosis
2. Right Ventricular Hypertrophy
3. Overriding Aorta
4. Ventricular Septal Defect
JB

- 26 year old man born with Tetralogy of Fallot.
- Surgical repair at 13 months old with trans-annular patch.
Tetralogy of Fallot
Surgical Management
TOF: Blalock-Taussig Shunt 1944 “Artificial Ductus Arteriosus”
TOF 1954: Complete Repair with VSD Patch, Trans-annular Patch, +/- Pulmonic Valvotomy, Infundibular Muscle Bundle Resection
• 26 year old man born with Tetralogy of Fallot.
• Surgical repair at 13 months old with trans-annular patch.
• Pulmonic valve replacement with homograft at 11 years old.
TOF Repaired with Trans-annular Patch: Chronic Severe Pulmonic Regurgitation
Chronic PR s/p TAP Repair of TOF: When to Replace the Pulmonic Valve?

• Moderate or greater PR + symptoms (RHF, decreased exercise tolerance) → PVR is indicated.

• However, irreversible RV dysfunction usually precedes symptoms.

• What metric tells us the latest time at which the asymptomatic patient should have PVR to prevent irreversible RV dysfunction?

• CMR study of 17 patients: no patient with RVEDVI >170 ml/m2 or RVESVI > 85 ml/m2 normalized their RV volumes after PVR (Therrien J, Am J Cardiol 2005).

• ACC 2018 ACHD Guidelines...
rTOF with TAP and Chronic PR (moderate or greater): Current Indications for PVR

• Symptoms (decreased exercise tolerance, right heart failure) or
• RVEDVI > 160 ml/m2 or RVESVI > 80 ml/m2 or RVEDV >2X LVEDV.
Assessment of RV Size

TTE challenging: retrosternal location, complex geometry.

CMR is accurate and reproducible.
(PVR threshold RVEDVI 160 ml/m2)
CMR: PR RF = retrograde flow/antegrade flow
### CMR Imaging

#### Recommendations for CMR Imaging

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<th>COR</th>
<th>LOE</th>
<th>Recommendations</th>
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<tr>
<td>I</td>
<td>B-NR</td>
<td>In patients with ACHD who have or who are at risk of developing RV enlargement and dysfunction, serial CMR is recommended for quantitative assessment of RV size and function.</td>
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<tr>
<td>Ila</td>
<td>C-LD</td>
<td>CMR can be useful in the initial evaluation and serial assessment of selected patients with CHD based on anatomic complexity and clinical status.</td>
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Surgical PVR for PR s/p Tet Repair (“Native RVOT”)

- Mean age 29 years old, range 7-40 years old.
- Surgical PVR is current standard of care. Percutaneous valve implantation is investigational (Medtronic Harmony Valve).
- Median time to replace the surgically placed valve is 10-15 years.
JB

• 26 year old man born with Tetralogy of Fallot.
• Surgical repair at 13 months old with trans-annular patch.
• Pulmonic valve replacement with homograft at 11 years old.
• Pulmonic valve replacement with 23 mm porcine bio-prosthesis at 15 years old.
Pulmonic Homograft Stenosis
Prone to Inflammatory/Fibrotic/Calcific Stenosis
Dysfunction of Surgically Place Pulmonic Valve

• Most common treatment for dysfunction of surgically placed valve is Percutaneous Pulmonic Valve Implantation (Medtronic Melody or Edwards Sapien) unless technically not feasible.
Percutaneous Pulmonic Valve Implantation: Treatment of Conduit Dysfunction in rTOF
Repaired Tetralogy of Fallot: Beyond the Pulmonic Valve

• Tricuspid regurgitation.
• Residual VSD.
• TAP Aneurysm
• Residual RVOT obstruction (infundibular, valvular, supravalvular).
• Aortic dilatation (mild dilatation is common, dissection is rare).
• Aortic regurgitation.
• LV dysfunction.
• Right sided aortic arch.
• Extracardiac RV-PA Conduit (pulmonary atresia, anomalous coronary artery).
• 26 year old man born with Tetralogy of Fallot.
• Surgical repair at 13 months old with trans-annular patch.
• Pulmonic valve replacement with homograft at 11 years old.
• Pulmonic valve replacement with 23 mm porcine bio-prosthesis at 15 years old.
• Balloon pulmonary valvuloplasty at 25 years old (PS moderate to severe pre procedure, moderate post procedure).
JB: Self refers to Keck...
Do I need a defibrillator?
"rTOF: Cardiac death can occur at any stage and structural re-intervention is common"

Congenital Heart Disease, Volume: 12, Issue: 3, Pages: 301-308, First published: 28 November 2016, DOI: (10.1111/chd.12439)
ACC ACHD 2018 TOF: ICD for “...standard qualifying criteria” (LVEF 35% + FC II/III, SCA, sustained/symptomatic VT). Otherwise IIa for multiple risk factors.

3. Programmed ventricular stimulation can be useful to risk-stratify adults with TOF and additional risk factors for SCD (S4.3.5-3–S4.3.5-8).

7. Risk factors for SCD include:
   a. LV systolic or diastolic dysfunction
   b. Nonsustained VT
   c. QRS duration ≥180 ms
   d. Extensive RV scarring
   e. Inducible sustained VT at electrophysiological study
JB: 26 years old, repaired TOF

• TTE: Normal LV systolic and diastolic function.
JB: 26 years old, repaired TOF
Surveillance monitor March 2018
JB: 26 years old, repaired TOF
Trans-annular Patch
Later Aneurysm
JB: Self refers to Keck…
Do I need a defibrillator?

7. Risk factors for SCD include:
   a. LV systolic or diastolic dysfunction
   b. Nonsustained VT
   c. QRS duration $\geq$ 180 ms
   d. Extensive RV scarring
   e. Inducible sustained VT at electrophysiological study

• EP Study appropriate.
JB: TOF, EP Study Positive: Referred for Primary Prevention ICD
TOF: Future Directions
TOF New Surgical Techniques: No TAP, Less PR
Finding the Ideal Marker to Indicate PVR for Chronic PR s/p TAP Repair of TOF
Chronic Aortic Regurgitation “Inflection Point”
Chronic PR Inflection Point??


- Rationale and design of the Canadian Outcomes Registry Late After Tetralogy of Fallot Repair: the CORRELATE study.
Is PPVI Non-inferior to Surgical PVR for Chronic PR s/p TAP Repair of TOF? (The Native RVOT)
Transcatheter Harmony Valve for Native RVOT: Two Year Outcomes from Early Feasibility Study

- n = 18
- No paravalvular leaks.
- Two patients experienced tissue growth within the stent frame and were successfully treated with Melody valve-in-valve.
- 40 patient Harmony Pivotal Trial currently enrolling.

- April, 2018 SCAI Scientific Sessions
Fontan (Functional Single Ventricle)
O.S.

• 22 year old man born with unbalanced atrioventricular septal defect with hypoplastic right ventricle, pulmonary atresia and L-TGA.
Atrioventricular Septum, Normal Heart
Normal Heart vs Partial AVSD (Primum ASD + Cleft Mitral Valve)
Complete AVSD: Primum ASD+ Inlet VSD, Common Atrio-Ventricular Valve. RV and PA Volume Overload
Surgical “Septation” of Complete AVSD
Unbalanced, “Un-septatable” AVSD

- Approximately 10-15% are unbalanced

- Left Dominant: ~1/3
- Right Dominant: ~2/3
O.S.: Unbalanced atrioventricular septal defect with hypoplastic right ventricle, pulmonary atresia and L-TGA

Pulmonary flow dependent of patent ductus arteriosus
O.S.

• 22 year old man born with unbalanced atrioventricular septal defect with hypoplastic right ventricle, pulmonary atresia and L-TGA.
• 4 days old: Blalock Taussig Shunt.
Modified Blalock Taussig Shunt
O.S.

• 22 year old man born with unbalanced atrioventricular septal defect with hypoplastic right ventricle, pulmonary atresia and L-TGA.
• 4 days old: Blalock Taussig Shunt.
• 6 months old: Bidirectional Glenn (SVC to RPA).
O.S.

- 22 year old man born with unbalanced atrioventricular septal defect with hypoplastic right ventricle, pulmonary atresia and L-TGA.
- 4 days old: Blalock Taussig Shunt.
- 6 months old: Bidirectional Glenn (SVC to RPA).
- 4 years old: Non-fenestrated, extracardiac Fontan.
Modern Fontan Circulation
Functional Single Ventricle
Fontan Circulation

A. Biventricular Circulation

LV

P_{Atrium} 5

Lungs

P_{PA} 15

RV

P_{IVC/SVC} 5

Body

B. Univentricular Fontan

SV

P_{Atrium} 2

Lungs

P_{PA} 12

Body

P_{IVC/SVC} 15
Fontan Univentricular Circulation

• Palliative, physiologic, non-anatomic repair.

• Systemic venous blood to lungs:
  Resolves cyanosis.
  Relieves volume overload of systemic ventricle.

• Chronic systemic venous congestion.
• Chronic low C.O. with limited reserve.
Hepatic Fibrosis Is Universal Following Fontan Operation, and Severity is Associated With Time From Surgery: A Liver Biopsy and Hemodynamic Study

David J. Goldberg, MD; Lea F. Surrey, MD; Andrew C. Glatz, MD, MSCE; Kathryn Dodds, CRNP; Michael L. O’Byrne, MD, MSCE; Henry C. Lin, MD; Mark Fogel, MD; Jonathan J. Rome, MD; Elizabeth B. Rand, MD; Pierre Russo, MD; Jack Rychik, MD
Fontan-Associated Liver Disease

Proceedings from the American College of Cardiology Stakeholders Meeting, October 1 to 2, 2015, Washington DC
The Fontan circulation: Sources of hypoxemia

• Veno-venous collaterals.
• Fontan fenestration.
A. Normal physiology

B. Collateral circulation

These shunting connections are 1) the aortopulmonary collaterals (between the bronchial artery and the pulmonary artery), 2) the veno-venous collaterals (between the bronchial vein and the pulmonary vein) and 3) the arterio-venous shunts (direct connections between the bronchial artery and vein bypassing the capillary network).
Veno-venous Collaterals: Left Brachiocephalic Vein to Pulmonary Veins (R to L shunt)
Fontan Fenestration

- Decrease pleural effusions post-op.
- Small R to L shunt (~2% drop in sat).
- Decrease hepatic congestion.
- Increase systemic output.
- Potential for access for ablation, hemodynamic measurements, pacemaker leads.
- Potential for paradoxical embolism.
- Test occlusion: fail if >3 mmHg increase in CVP or drop in BP.
Atrial Arrhythmias in Fontan Patients

- ~50% lifetime prevalence of atrial arrhythmias.
- Atrial arrhythmias commonly precipitate overt heart failure.
- Prompt TEE guided DCCV generally recommended.
- Anti-coagulate.
- Consider ablation.

![ECG waveform](image)
Intracardiac Thrombus in Fontan Patients

• Risk factors for development: low flow, atrial arrhythmias, prosthetic material, blind pouches, hypercoagulability.
• ~30% lifetime prevalence.
• High morbidity and mortality.
Fontan: Chronic Management

• Anticoagulation for atrial arrhythmia or thromboembolic event. Evidence accumulating that NOACs are acceptable.
• Conventional heart failure therapies: no conclusive evidence.
• PDE-inhibitors and endothelin receptor antagonists improve hemodynamics and functional capacity and are indicated for symptomatic patients.
• Screen for HCC, ensure Hep A and B immunity, and check for active hepatitis C.
• Walk and exercise!!!
• Consider heart +/- liver transplant (many challenges).
“Exercise training is safe and beneficial...”

Heart, Lung and Circulation 2015
Nigel Sutherland, BPhysio

- “Muscle Pump” (one way valves in veins in muscles).
- “Ventilatory Pump” (negative intrathoracic pressure augments venous return).
- Increased muscle mass.
- Peripheral conditioning (eg O2 dissociation).
- Improved functional capacity and quality of life.
Fontan: Acute Management
CTPA for Diagnosis of PE in Fontan Patients

• False positives due to streaming artifact.
• False negatives due to incomplete filling.
• Increasing contrast volume, increasing injection rate and delaying acquisition may improve accuracy; discuss with radiologist.
Inotropes and pressors have no effect unless systemic ventricular dysfunction predominates.
Avoid Nitrates and Over-diuresis. Caution with Positive Pressure Ventilation

• “Congestive Force” is required to maintain filling of the systemic ventricle.
Fontan Future Directions: Is there a solution?
AHCD Take Homes

• The ACHD population continues to grow.
• ACHD fellowship training, board certification and center accreditation are established.
• Repaired TOF research priorities: optimal timing and method for PVR, accurate risk assessment for SCD.
• Fontan research priorities: augment flow away from the viscera and into the pulmonary arteries, standardize selection for transplant.
• Fontan: avoid nitrates; caution with diuresis and positive pressure ventilation.
Dr. Roberta Williams

• “Congenital heart disease is an infinite combination of structural and physiologic variables.”