Adult Congenital Heart Disease: A Growing Problem

Dr. Gary Webb
Cincinnati Children’s Hospital Heart Institute
## ACHD Resources in Ohio

<table>
<thead>
<tr>
<th>City</th>
<th>State/Prov.</th>
<th>Institution</th>
<th>Medical Director</th>
<th>Patient Vists/Yr.*</th>
<th>ACHD Oper/Yr.**</th>
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<td>Akron</td>
<td>OH</td>
<td>Adult Congenital Heart Service</td>
<td>John R. Lane, MD</td>
<td>379</td>
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<td>Summa Health System, Akron General Medical Center</td>
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<td>(330) 543-8521</td>
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<tr>
<td>Cincinnati</td>
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<td>Adult Congenital Heart Disease Clinic</td>
<td>Gary Webb, MD</td>
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<td>Cincinnati Children’s Hospital Medical Center</td>
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<td></td>
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<td>(513) 636-7593</td>
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<td>Cleveland</td>
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<td>Cleveland Clinic Adult Congenital Heart Program</td>
<td>Richard Krasuski, MD</td>
<td>824</td>
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<td>Cleveland Clinic</td>
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<td>(216) 445-7430</td>
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<tr>
<td>Columbus</td>
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<td>COACH (Columbus Ohio Adult Congenital Heart) Program</td>
<td>Curt J. Daniels, MD</td>
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<td></td>
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<td>Nationwide Children’s Hospital</td>
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<td>(614) 722-5622; (614) 293-6638</td>
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<tr>
<td>Cleveland</td>
<td>OH</td>
<td>ADCON, Congenital Heart Center, Heart &amp; Vascular Institute</td>
<td>Ernest Siwik, MD</td>
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<td>Rainbow Babies &amp; Children’s Hospital, University Hospitals</td>
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<td>(216) 844-7700</td>
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Situations When We Might Help

- When you don’t know a lot about a CHD diagnosis.
- When a CHD diagnosis includes a person’s name.
  - Fontan.
  - Mustard.
  - Senning.
  - Rastelli.
  - Jatene.
Situations When We Might Help

- CHD patients who are cyanotic.
- CHD patients with pulmonary hypertension.
- CHD patients with only one ventricle.
- CHD patients needing noncardiac surgery.
- CHD patients needing special cardiac surgery.
  - David operation.
  - Ross procedure.
  - Aortic valve repair.
Situations When We Might Help

- CHD patient candidates for a heart catheterization.
- Sexually active CHD patients.
- CHD patients contemplating pregnancy.
- Pregnant CHD patients.
- CHD patients with arrhythmias.
- CHD patients with syndromes.
Consider ACHD clinics to contain resource people who have developed networks of specialists with special interests and abilities.
The Toronto Congenital Cardiac Centre for Adults – founded 1959
Patient 63 - History

- 31-year-old female supermarket clerk
- 16 weeks pregnant.
- Cyanotic at birth.
- Left BT shunt at age 11.
- Lateral tunnel Fontan procedure at age 20.
- Lost to follow-up until age 31.
Patient 63 - History

- NYHA class I.
- No functional decline recently.
- No medications.
Patient 63 “Risk Factors”

- “Don’t know much about that”.
  - Fontan.
  - Blalock Taussig.
- Man’s name.
- Pregnancy.
What is a Fontan procedure?

- I don’t know.
- A type of transposition repair.
- A type of tetralogy repair.
- A type of single ventricle repair.
What are the pregnancy issues associated with a Fontan procedure?

Choose as many as are applicable

- Atrial flutter.
- Thromboembolism.
- Fetal loss.
- Cyanosis.
- Maternal death.
- All of the above.
Fontan Pregnancy Management

• Close monitoring.
• High-risk pregnancy team.
• Consider anticoagulation.
• Planned vaginal delivery.
• Planned epidural.
Patient 63
Patient Course and Outcome

- Induced at 39 weeks.
- Epidural anesthetic.
- Normal vaginal delivery.
- Healthy baby boy.
Patient 61 - History

- 24-year-old man, works as an engineer.
- Born with tricuspid atresia.
- Palliated with bilateral Blalock-Taussig shunts.
- “Atriopulmonary Fontan” procedure at age 6.
- Started on warfarin at age 16 for possible pulmonary embolism.
Patient 61 - History

- At age 17, developed recurrent atrial flutter.
- Treated with various antiarrhythmics, including amiodarone.
- By age 21, was in chronic atrial fibrillation.
- Was otherwise doing well (NYHA I).
Patient 61 - Physical Examination

- BP 118/72
- Pulse 70 (irregular)
- Height 68 inches
- Weight 137 pounds
- Pulse oximetry 96%
Patient 61 - Physical Examination

- Bilateral thoracotomy and midline sternotomy scars.
- JVP hard to assess.
- Chest clear.
- Normal precordial examination.
- Normal heart sounds with no murmurs.
- No hepatomegaly, ascites, or edema.
Patient 61 “Risk Factors”

- “Don’t know much about that”.
  - Fontan.
  - Blalock Taussig.
- Man’s name.
- Arrhythmia.
- One ventricle.
Patient 61 - What next steps?

- Optimize anticoagulation.
- Refer for immediate surgery.
- Consider thrombolysis.
- No specific treatment needed.
Patient 61
Decision and Management

• A Fontan revision was elected.
  – Right atrial thrombus removed along with excess right atrial tissue.
  – “Extracardiac Fontan” constructed.
  – Arrhythmia surgery not possible because of operative difficulties.
Patient 61
Decision and Management

- Immediately postoperatively
  - Hemodynamically unstable
  - Persistent bleeding
  - Sternal closure impossible, so skin only closure.
  - Maximum pressors.
Patient 61
Decision and Management

- Postoperative complications
  - Compartment syndrome left leg.
  - Recurrent tension pneumothorax.
  - Acute tubular necrosis
  - Tracheotomy..
  - Discharged home after four months, in atrial fibrillation, and on warfarin.
Patient 53 - History

• 38-year-old woman
• Original CHD diagnoses
  – TGA
  – VSD
  – PS
• Age 7, Rastelli procedure with a 20 mm Dacron conduit including a Hancock valve
Rastelli Procedure
Patient 53 - History

- Age 25, Strep sanguis endocarditis
- She was found to have severe conduit obstruction, moderate subaortic stenosis, a small residual VSD, and preserved biventricular function.
- Her conduit was replaced using a homograft, her VSD was enlarged to reduce the LVOT gradient, and a baffle was placed between LV and the aorta.
Patient 53 - History

- Age 35, heart catheterization showed a 40 mmHg gradient across the RV-PA conduit with mild LVOTO and mild AR. Sotalol was started for suspected paroxysmal atrial tachycardia.
- Age 38, NYHA class III with exertional dyspnea and fatigue. She is limited to 100 yards on the flat. She avoids steps.
- Current medications: warfarin; sotalol 80 mg BID; furosemide 20 mg daily; sertraline 100 mg daily
Patient 53 - Physical Examination

- BP 110/70
- Pulse 46
- Height 70 inches
- Weight 209 pounds
- Pulse oximetry 97%
Patient 53 - Physical Examination

- JVP 8 cm ASA.
- Chest clear.
- Possible right ventricular lift.
- Second sound loud and single.
- Grade 4 long ejection systolic murmur upper left sternal border.
- No hepatomegaly or edema.
How much conduit obstruction does she have?

- Mild.
- Moderate.
- Severe.
- Don’t know.
Patient 53 - Echo Report

- Normal LV size and function.
- Peak LVOT gradient 20 mmHg.
- RVH with mild-moderate hypokinesis.
- Mild TR.
- Valved conduit not seen.
Patient 53 – “Risk Factors”

- “Don’t know much about that”.
  - TGA.
  - VSD/PS.
- Man’s name.
- Exercise intolerance.
- Sexually active.
Patient 53
Patient
53
Patient 53
Why is she NYHA class III?

Choose as many as are applicable

- The stenosed conduit?
- The sotalol?
- Chronotropic incompetence?
- Anemia?
- Lung disease?
Patient 53 – Management and Outcome

- Conduit from RV to PA removed and replaced.
- Smooth postop course.
- Patient NYHA I after convalescence.
Patient 41 - History

• 31-year-old woman
• Original CHD diagnoses
  – Tetralogy of Fallot
• Age 3, transannular patch repair.
• Progressive pulmonary regurgitation, severe.
• Asymptomatic and full time work, but has “slowed down” a bit.
Patient 41 - Physical Examination

- BP 110/70
- Pulse 70
- Height 62 inches
- Weight 189 pounds
- Pulse oximetry 100%
Patient 41 - Physical Examination

- Midline sternotomy scar.
- Normal JVP.
- RV heave.
- Grade 2 ejection systolic murmur.
- Grade 2 low pitched diastolic murmur.
Patient 41 - Echo Report

- Normal LV size and function.
- Right ventricle severely dilated.
- Laminar (severe) pulmonary regurgitation.
Patient 41 – “Risk Factors”

- “Don’t know much about that”.
  - Tetralogy.
  - Pulmonary regurgitation.
- Man’s name.
- Candidate for a heart catheterization.
Patient
41
Patient 41
### Ventricular Volume Quantification

<table>
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<th>LV</th>
<th>(Normal range)</th>
<th>RV</th>
<th>(Normal range*)</th>
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<tbody>
<tr>
<td>EDV (mL)</td>
<td>116</td>
<td>(52–141)</td>
<td>244</td>
<td>(58–154)</td>
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<tr>
<td>ESV (mL)</td>
<td>32</td>
<td>(13–51)</td>
<td>88</td>
<td>(12–68)</td>
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<td>SV (mL)</td>
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<td>(33–97)</td>
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<td>(57–75)</td>
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<td>Mass index (g/m²)</td>
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<td>(47–77)</td>
<td>49</td>
<td>(20–40)</td>
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*Normal values based on gender, age, and body surface area.

Patient 41

Figure 41-6 Phase contrast velocity mapping of the main pulmonary artery.
Patient 41

Treatment Options?

Choose as many as are applicable

- Bioprosthetic pulmonary valve replacement.
- Mechanical pulmonary valve replacement.
- Percutaneous pulmonary valve replacement.
- Continued observation.
Patient 41 – Plan and Outcome

• Pulmonary valve replacement elected.
• 23 mm homograft used (bioprosthesis).
• Previous patch excised.
• Exercise intolerance for improved after convalescence.
• RV volumes reduced by 30% on follow-up MRI.
Patient 25 - History

- 45-year-old man
- Original CHD diagnoses
  - Aortic coarctation (isolated)
- Age 3, coarctation repair (details unknown).
- Recently found to have high blood pressure.
- Treated with an ACE inhibitor and a beta blocker.
- No cardiovascular symptoms.
Patient 25 - Physical Examination

- BP 156/70 right arm
  115/60 left arm
  118/? right thigh
- Pulse 44
- Height 70 inches
- Weight 202 pounds
- Pulse oximetry 98%
Patient 25 - Physical Examination

- Left thoracotomy scar.
- Pulse delay right radial to right femoral.
- Very weak pedal pulses.
- Grade 2 ejection systolic murmur.
- Grade 2-3 murmur of aortic regurgitation.
- Faint continuous murmur heard over the back.
Patient 25
Patient 25

Echo Report

- Bicuspid aortic valve.
- Mild aortic regurgitation.
- Normal left ventricular size and function.
- Coarctation gradient 47 mmHg with diastolic tail.
Patient 25 - MRI Report

- Mild dilation of the ascending aorta.
- Hypoplastic distal aortic arch with severe coarctation.
- Collateralization present.
Patient 25 - MRI
Patient 25 – “Risk Factors”

- Possible candidate for cardiovascular surgery.
- Possible candidate for a heart catheterization.
Patient 25
Treatment Options?
Choose as many as are applicable

- Surgical repair of aortic coarctation.
- Balloon dilation of aortic coarctation.
- Stent placement for aortic coarctation.
- More aggressive antihypertensive therapy.
Patient 25 – Plan and Outcome

- Stent therapy of aortic recoarctation elected.
## Patient 25 – Plan and Outcome

<table>
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<th>BP right arm</th>
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<td><strong>Prior to procedure</strong></td>
<td>156/70</td>
<td>115/60</td>
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<td><strong>3 weeks after procedure</strong></td>
<td>108/65</td>
<td>113/70</td>
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Case 19
Patient 19 - History

- 26-year-old woman
- Original CHD diagnoses
  - Normal heart
  - Infective endocarditis
- Age 9 and 10, aortic valve repair for aortic regurgitation.
- Age 18, Ross procedure.
- Age 26, declining exercise capacity, limited to two floors vertically.
Patient 19 - Physical Examination

- BP 100/42
- Pulse 100
- Height 70 inches
- Weight 147 pounds
- Pulse oximetry 100%
Patient 19 - Physical Examination

- Bounding peripheral pulses.
- Positive Duroziez sign.
- Cardiac apex displaced and active.
- Grade 2 medium length ejection systolic murmur.
- Grade 2 high-pitched diastolic murmur LSB.
Patient 19 - Echo Report

- Left ventricle dilated to 62 mm diastolic.
- Normal LV systolic function.
- Ascending aortic dilation to 55 mm.
- Severe neoaortic valve regurgitation.
- Homograft function normal on the right side of the heart.
Patient 19 - Echo
Patient 19 - Echo
Patient 19 – “Risk Factors”

- CHD patients needing special cardiac surgery.
  - David operation.
  - Ross procedure.
  - Aortic valve repair?
Patient 19 - Which management would you consider?

Choose as many as are applicable

- Bentall procedure with mechanical aortic valve.
- Bentall procedure with bioprosthetic aortic valve.
- Valve sparing root replacement.
Patient 19 – Management and Outcome

- Valve sparing aortic root replacement elected.
- At surgery, the neoaortic valve leaflets appeared normal.
- The aortic annulus was less dilated than were the sinuses.
- The aorta was replaced using a David technique, sewing a 26 mm Dacron graft with preformed sinuses down to the annulus with resuspension of the neoaortic leaflets.
- The coronary arteries were reimplanted.
Patient 19 – Management and Outcome

- The patient did well with no residual aortic valve regurgitation.
Patient 19 – Management and Outcome
Patient 6 - History

- 34-year-old woman
- Generally healthy childhood.
- Told that her heart was on the right side.
- Occasional episodes of hemoptysis as a teenager.
- Hemoptysis recurred after a recent successful pregnancy.
- Lifelong non-smoker.
- Very good exercise capacity.
Patient 6 - Physical Examination

- BP 100/68
- Pulse 62
- Height 56 inches
- Weight 141 pounds
- Pulse oximetry 98%
Patient 6 - Physical Examination

- Normal JVP.
- Clear chest.
- Right-sided apical impulse.
- Heart sounds normal, no murmurs.
- Right-sided liver.
Patient 6 - Echo Report

- Dextroposition of the heart with normal cardiac chamber anatomy.
- Mild right heart dilation.
- Normal RVSP.
## Patient 6 - MRI

### Ventricular Volume Quantification

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Patient 6 - CT
Patient 6 – “Risk Factors”

• “Don’t know much about that”.
  – Anomalous pulmonary venous connection.
  – Scimitar syndrome.
Patient 6 - What management would you consider?

Choose as many as are applicable

- Observation.
- Surgical repair of scimitar circulation.
- Embolization of anomalous arterial supply.
- Surgical resection of abnormal lung segment.
Patient 6 – Management and Outcome

- Observation was elected.
- Many patients with scimitar syndrome have a shunt less than 2/1 and do not need redirection of their venous return.
- She did not have lung sequestration on CT scanning. Her lung parenchyma was normal.
- Over an 8-year follow-up, the patient has done well.