Management of Heart Failure in Adult with Congenital Heart Disease

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Background

• 0.4% of adults have some form of CHD
• 0.04% - 0.14% of adults have severe CHD (depending on geographic region and definition of severe)

The number of patients with severe CHD that will be presenting with emergent conditions is growing

Marelli et al, Circ, 2007;115;163-172
• Surgery is the cornerstone in the management of complex congenital heart disease
• 90% of patients with CHD will survive to adulthood instead of 20% (pre-surgical era)

• Complete repair is extremely unlikely and most patients with complex CHD are considered palliated
• Majority of patients are left with residual anatomical or hemodynamic abnormalities that predispose them to HF
How do patients with complex CHD present?

- Arrhythmia
- HF
- Stroke
- IE
When do patients with Complex CHD present with HF?

<table>
<thead>
<tr>
<th>Congenital Heart Diagnosis</th>
<th>Age during Non-Heart Failure-Related Hospitalization (Years, 95% CI)</th>
<th>Age during Heart Failure-Related Hospitalization (Years, 95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Atrial septal defect</td>
<td>57.4 (56.7–58.2)</td>
<td>68.0 (66.7–69.2)</td>
</tr>
<tr>
<td>Bicuspid aortic valve/congenital aortic insufficiency</td>
<td>50.7 (49.6–51.7)</td>
<td>58.7 (57.0–60.4)</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>43.5 (42.0–44.9)</td>
<td>55.9 (53.6–58.2)</td>
</tr>
<tr>
<td>Congenital coronary anomaly</td>
<td>56.2 (55.2–57.2)</td>
<td>63.9 (61.3–66.6)</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>34.8 (32.8–36.8)</td>
<td>45.0 (41.1–49.0)</td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>51.1 (47.4–54.7)</td>
<td>66.4 (60.9–72.0)</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>45.3 (41.4–49.2)</td>
<td>58.0 (52.8–63.3)</td>
</tr>
<tr>
<td>Congenital aortic stenosis</td>
<td>45.1 (41.9–48.4)</td>
<td>54.3 (48.0–60.6)</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>41.6 (38.9–44.3)</td>
<td>54.1 (49.1–59.1)</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>38.5 (33.5–43.4)</td>
<td>43.9 (34.0–53.8)</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>34.7 (30.7–38.7)</td>
<td>44.1 (36.2–52.0)</td>
</tr>
</tbody>
</table>

83000 ACHD patients admitted in USA in 2007
How do patients with complex CHD die?
Mechanism of HF

- Chronic volume and/or pressure overload
- Inadequate myocardial preservation during prior surgery
- Myocardial fibrosis
- Surgical injury to coronary artery
- Pulmonary hypertension
As adults with complex CHD get older, acquired heart disease and its risk factors become more frequent and contribute to the development of HF.

HF in ACHD patients are less likely to be due to LV systolic dysfunction as in patients with acquired HF.
HF in Congenital Heart Disease

Bimodal Classification

- Systolic Dysfunction
- Diastolic Dysfunction

- Left Heart
- Right Heart
- Single Heart
Risk factors for CAD in ACHD’s patient

• Congenital coronary abnormalities (anomalous origin &/or course)
• Iatrogenic injury during previous repair
• Intentional manipulation during previous surgical repair such as re-implantation of coronary artery during arterial switch or root replacement in Marfan patient
• Atherosclerosis
• All traditional risk factors associated with atherosclerosis are present in considerable number of ACHD patients
• Among 1976 ACHD patients, 82% were found to have >=1 RF
  • HTN 52%
  • Dyslipidemia 25%

• 40% of ACHD patients are overweight or obese
• Lack of exercise or wrong prohibitive advice

American Journal of Cardiology 2012; 109: 1657
Evaluation

• **Absolute** understanding or underlying anatomy and previous surgeries, repairs and intervention

• Look for anatomical dysfunction:
  1. Residual shunts
  2. Baffle stenosis
  3. Valvular dysfunction
  4. Conduit dysfunction
  5. Collaterals vessels

• Look for arrhythmia
Clinical Presentation Related to Coronary Artery Disease.

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>N = 141</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (%)</td>
</tr>
<tr>
<td>Initial Clinical Presentation</td>
<td></td>
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<tr>
<td>Angina pectoris</td>
<td>49 (35%)</td>
</tr>
<tr>
<td>Myocardial infarction</td>
<td>39 (27%)</td>
</tr>
<tr>
<td>Asymptomatic</td>
<td>53 (38%)</td>
</tr>
<tr>
<td>Indication for angiogram</td>
<td></td>
</tr>
<tr>
<td>Assessment of coronary arteries</td>
<td>86 (61%)</td>
</tr>
<tr>
<td>Other*</td>
<td>55 (39%)</td>
</tr>
<tr>
<td>Severity of Disease</td>
<td></td>
</tr>
<tr>
<td>One vessel disease</td>
<td>60 (43%)</td>
</tr>
<tr>
<td>Two vessel disease</td>
<td>36 (25%)</td>
</tr>
<tr>
<td>Three vessel disease</td>
<td>31 (22%)</td>
</tr>
<tr>
<td>Left main CAD</td>
<td>14 (10%)</td>
</tr>
<tr>
<td>Coronary Interventions</td>
<td></td>
</tr>
<tr>
<td>PCI alone</td>
<td>23 (16%)</td>
</tr>
<tr>
<td>PCI combined with CHD surgery</td>
<td>2 (1%)</td>
</tr>
<tr>
<td>CABG alone</td>
<td>18 (13%)</td>
</tr>
<tr>
<td>CABG combined with CHD surgery</td>
<td>66 (47%)</td>
</tr>
</tbody>
</table>

14% with CAD were less than 40 years (premature)
HF Management

Pump Model

- HF is caused by dysfunction of one or both pumps.
- Treatment is directed to improve the pump function or decrease the resistance that is acting against the pump.

Neurohormonal and cardiac remodeling Model

Heart Failure in Children and young adult
Ed 1, 2006
Management of ACHD patients with LV dysfunction

• Identification and relief of residual obstruction and/or regurgitation

• Conventional therapy may have some rule in ACHD patients with normal connections and no residual correctable lesion
Complex CHD

- Repaired TOF
- Systemic RV
- Fontan Patients
Tetralogy of Fallot (TOF or "Tet")

- Mitral Valve
- Aorta Shifting to Right
- Opening Between Ventricles
- Tricuspid Valve
- Pulmonary Valve
- Right Ventricular Outflow Obstruction

AO = Aorta
PA = Pulmonary Artery
LA = Left Atrium
RA = Right Atrium
LV = Left Ventricle
RV = Right Ventricle

- Red = Oxygen-rich Blood
- Blue = Oxygen-poor Blood
- Purple = Mixed Blood
Medical Treatment Trials in TOF (post surgery)

BB

33 patients
6 mo
Bisoprolol vs. placebo
NYHA I-II
BNP > 100
Vo2 max <25

ACEI

64 patients
6 mo
Ramipril Vs. Placebo

International Journal of Cardiology 154 (2012) 299–305
Systemic Right Ventricle
TGA → → → Mustard-Senning

The great arteries arise from the incorrect ventricle

The morphologic right ventricle acts as the systemic ventricle
CC-TGA

The morphologic right ventricle is in the systemic position.
ACEI in systemic RV

17 patients

Int J Cardiolog 2008; 129: 187
ARB in systemic RV

88 patients
The whole cohort did not show difference but symptomatic patient had an improvement in RV EF%
Valsartan as an example

**ACHD**
- In Van der Bom et al study
- The plan was to use multicenter to include 128 patients
- Only 88 patients were enrolled (out of 323 eligible patients)
- Only 62 patients ended the study
- The largest and best study of its kind

**Normal structure acquired HF**
- Val-HeFt trial
- 5010 patients were included

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We needed 102 patients for 80% power to detect a difference in EF of 5%. Possibly, Underpowered.
BB in systemic RV

• Some small studies showed some benefit
• Be carful, since many of these patients have an underlining sinus node dysfunction
So beside statics, what else can explain the failure of these medical management?

- Incomplete understanding of pathophysiology

- In patients with repaired TGA (Mustard or Senning), failure of atrio-ventricular coupling + inability to increase the stroke volume with dobutamin stress $\rightarrow$ limit stroke volume and COP responds (it is an inherited baffle problem)

- In cc-TGA, it is the valvular disorder and patients who stay free from significant TR (systemic AV valve) are free of HF for decades
Fontan Anatomies

Fontan Procedure
For heart with only one usable ventricle
(in this illustration Tricuspid Atresia)

1. AO
2. PA
3. LA
4. RA
5. LV

- stitches
- artificial wall to make chamber
- inferior vena cava
• In Patient post Fontan palliation, there is no sub-pulmonic ventricle
• Pulmonary vascular resistance is the major determinant of circulatory output
• Ventricular function is important only when it is severely impaired
- Small studies of pulmonary vasodilator showed some promising result
- However, larger and better designed studies are needed
Other Therapy

• ICD
• CRT
• Heart Transplant
• Mechanical device
What really work?
Patients referred to ACHD centres (per 1000 ACHD population per year)

Deaths (per 1000 ACHD population per year)

Year


+1.4%; P = 0.07

+2.0%; P = 0.24

+7.4%; P < 0.0001

Interaction P < 0.0001

Guideline presentation

Guideline publication

Change-point visits

Change-point deaths

-5.0%; P = 0.06

Guideline publication

Interaction P = 0.04
Summary

• HF in ACHD patients are less likely to be due to LV systolic dysfunction as in patients with acquired HF
• It is usually due to RV dysfunction, valve dysfunction or shunting
• Conventional HF therapy may have a rule in symptomatic patient
• HF in ACHD patients carries worse survival
• Large and well designed trials are needed
• Large and well designed trials are very difficult to be done
• Referral to specialized center is crucial for these patient’s wellbeing and survival
• Conventional RF modifications are required in all patients
Thank you