Heart Failure Themes: Congenital Heart Disease

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No Conflicts of Interest

One team shared values

East Midlands Congenital Heart Centre
The normal heart: anatomy
The normal heart: anatomy
The normal heart: function and flow
The normal heart: function and flow
Descriptions of healthy physiology define the normal heart and circulation

- Anatomy
- Function
- Pressure
- Flow
- Exercise physiology
- BNP
- Neurohormones
- Cytokines
- Haematological variables
We use those same variables to describe heart failure
As the resolution of those measurement techniques increases, we can recognise smaller abnormalities of physiological and anatomical variables and thereby diagnose heart failure at an earlier and earlier stage.
Hippocrates (c. 467-377BC)
- symptoms and signs of HF
- the heart was literally failing

21st century
- echo
- BNP
- aldosterone
- Na
- epinephrine
- norepinephrine
- CPEX
- cath
- Renin
- creatinine
- MRI
- CXR
- Hb

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The definition of heart failure?

...the description of the abnormal heart
Time line to an abnormal heart

Fetal heart failure  CHD  HCM  DCM  Myocardial infarction  Aortic stenosis  Senescence

AGE

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Heart failure (an abnormal heart) is not a final common pathway but a common beginning.
Congenital heart disease is an abnormal heart from birth

Those with congenital heart disease are born with heart failure
Form, function and flow are abnormal from the beginning
NHs are elevated in diverse anatomical groups
...and predict outcome

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Exercise capacity is truncated

Diller et al. Circulation 2005

ANOVA P<0.0001
...and predicts survival
NH activation and exercise limitation equivalent to patients with LV dysfunction

**A.**

- **MVO₂ (ml/kg/min)**
- **VE/VCO₂**

**B.**

- **Norepinephrine (nmol/L)**
- **Epinephrine (nmol/L)**
Case: 21 yr old male

- Paediatric clinics until 5yrs of age with “slow heart beat”
- Well through school years
- Working as a cleaner
- Gave up job at East Midlands Airport: unsociable hours
- Worsening exercise capacity Dec 2012
- Community access echo Feb 2013
- ACHD OPD March 2013: NYHA II/III
Case: 21 yr old male

- Cachectic: hgt 172cm, wgt 54kg, BMI 18.2kg/m²
- HR 50/min
- BP 124/58
- Apex: MAL
- 4/6 PSM and split S2
- Clear lung fields
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Diagnosis: congenitally corrected transposition of the great arteries

- Systemic right ventricle
- Sub-aortic AV valve = tricuspid valve
- Propensity to heart block
We know there is heart failure
How bad is it?
Creatinine 85
Hb 13.8
BNP 885ng/L
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Systemic RVEF 35%

One team shared values
### Condition 1: Air

<table>
<thead>
<tr>
<th>Vessel</th>
<th>Saturations</th>
<th>Pressures</th>
</tr>
</thead>
<tbody>
<tr>
<td>IVC</td>
<td>34.8%</td>
<td>Mean 15mmHg</td>
</tr>
<tr>
<td>SVC</td>
<td>31.1%</td>
<td>Mean 19mmHg</td>
</tr>
<tr>
<td>RA</td>
<td>35.9%</td>
<td>Mean 16mmHg</td>
</tr>
<tr>
<td>Subpulmonary ventricle (LV)</td>
<td>48.7%</td>
<td>130/74, mean 96 mmHg</td>
</tr>
<tr>
<td>Pulmonary artery (left)</td>
<td></td>
<td>128/10, EDP 19mmHg</td>
</tr>
<tr>
<td>LPA wedge</td>
<td></td>
<td>40mmHg</td>
</tr>
<tr>
<td>Aorta</td>
<td>94.2%</td>
<td>115/69, mean 87mmHg</td>
</tr>
<tr>
<td>Subsystemic ventricle (RV)</td>
<td>94.2%</td>
<td>103/5, mean 16mmHg</td>
</tr>
</tbody>
</table>

### Calculations:
- Assumed Vo2: 202.2 l/min
- Hb: 12.4g/dl
- Qp: 2.25l/min
- PVR: 24.7 WU
- PVR: 15.6 WU/m2
- SVR: 30.9 WU (hand calc 27.3)
- CO: 2.27l/min (hand calculated 2.6 l/min though)
- Transpulmonary gradient: 56mmHg

### Condition 2: 30PPM NO in air

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<tr>
<td>RA</td>
<td>17 mmHg</td>
<td></td>
</tr>
<tr>
<td>Subpulmonary ventricle (LV)</td>
<td>33.5%</td>
<td>130/75, mean 97mmHg</td>
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<tr>
<td>Pulmonary artery (left)</td>
<td>34.6%</td>
<td>130/75, EDP 25mmHg</td>
</tr>
<tr>
<td>LPA wedge</td>
<td></td>
<td>48mmHg</td>
</tr>
<tr>
<td>Aorta</td>
<td>92%</td>
<td>109/70, mean 80mmHg</td>
</tr>
<tr>
<td>Subsystemic ventricle (RV)</td>
<td>33.5%</td>
<td>112/14, mean 25mmHg</td>
</tr>
</tbody>
</table>

- PAP: 130/75/97
- TPG: 50mHg
- PVR: 24WU
- CO: 2.3L/min
- No reversibility with NO

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One team shared values
We know there is heart failure
How should we treat it?
The same principles apply

The same evidence is not there
• Ramipril
• Warfarin
• Bisoprolol
• Loop and thiazide diuretics
• Spironolactone → eplerenone
• Intravenous iron
• Specialist ACHD nurse input

• Psychological and social support for patient and family
Transplant assessment (Newcastle): turned down
• Steady deterioration
• NYHA III then IV
• Liver congestion and capsular pain
• Intractable pleural effusions
• Deteriorating renal function
Worsening RV function and TR

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• Admitted from clinic November 2014

• PEA arrest
Learning points

• Heart failure exists from birth in those with congenital heart disease

• Life long follow is critical: from the start of life

• HF can be quantified by conventional means

• Treatment approach is extrapolated from acquired HF but is not evidence based

• HF can be lethal at a young age in CHD

• The structure of care for ACHD should incorporate HF pathways and vice versa

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