Liver and the Circulation
From Galen to Fontan
Déjà vu all over again

William Evans
Professor Pediatrics University of Nevada
Disclosures

- No conflicts of interest
History of the Circulation

- 2,000 years of history
- < 15 minutes
- Frescos: La Historia de la Cardiología by Diego Rivera
  - National Institute of Cardiology Mexico City 1943
1st century AD
Galen

- Lived around 120-200 AD
- Greek but spent most of his time in Rome
- Physician to Roman emperors and the gladiators
Gap

- Roman Empire collapsed
- European Dark Ages - 1,000 years
- Renaissance
16th century AD
Andres Vesalius

- Born in Brussels in 1514
- Physician to Habsburgs, especially Charles V
  - Charles V sent the conquistadores to South America
- Professor anatomy University of Padua 29 years old
- Direct anatomic dissection & went against Galen
- Published *Humani corporis fabrica libri septem*-1543
Cy commence le m'liure des glats des m. clemens z'des m. humeur.
WILLIAM HARVEY (1578-1657)
William Harvey

- Born in England 1578
- Educated at Cambridge and Padua
- Court of Charles I
- Used experimental data ... voilà ... the circulation
On the Motion of the Heart and Blood - 1628
Single Ventricle Heart

- Maude Abbott publication ~1900
- Stimulated her interest in CHD
- Stymied early heart surgery
Surgical repair of tricuspid atresia

F. Fontan and E. Baudet

Centre de Cardiologie, Université de Bordeaux II, Hôpital du Tondu, Bordeaux, France

Surgical repair of tricuspid atresia has been carried out in three patients; two of these operations have been successful. A new surgical procedure has been used which transmits the whole vena caval blood to the lungs, while only oxygenated blood returns to the left heart. The right atrium is, in this way, 'ventriclized', to direct the inferior vena caval blood to the left lung, the right pulmonary artery receiving the superior vena caval blood through a cava-pulmonary anastomosis. This technique depends on the size of the pulmonary arteries, which must be large enough and at sufficiently low pressure to allow a cava-pulmonary anastomosis. The indications for this procedure apply only to children sufficiently well developed. Younger children or those whose pulmonary arteries are too small should be treated by palliative surgical procedures.
Fontan Procedure for HLHS

Superior vena cava connected to pulmonary artery

Tube outside heart

Inferior vena cava connected to tube
Fontan

~ 13-15 mm Hg

~ 0-5 mm Hg
Liver Blown Up

Fontan pressure
Single Ventricles

Right ventricles

Left ventricles
Single Ventricles and Pulmonary Blood flow at Birth

- Single ventricles
  - LV & *un*obstructed pulmonary blood flow
  - RV & *un*obstructed pulmonary blood flow
  - LV & *ob*structed pulmonary blood flow
  - RV & *ob*structed pulmonary blood flow
Fontan Outcomes and Pulmonary Blood Flow at Birth

William N. Evans¹,² · Ruben J. Acherman¹,² · Leigh C. Reardon¹,³ · Michael L. Ciccol¹,² · Alvaro Galindo¹,² · Abraham Rothman¹,² · Brody J. Winn⁴ · Noel S. Yumiaço⁴ · Humberto Restrepo¹,²

Received: 24 March 2015 / Accepted: 9 July 2015
© Springer Science+Business Media New York 2015
Fontan Outcomes and Pulmonary Blood Flow at Birth

- Pediatric Cardiology 2015
- 200 patients: 56 underwent transvenous-hepatic biopsy
- Bx average total score (pf 0-4+sf 0-4) for the 4 groups
  - (18%) LV & unobstructed pulmonary blood flow: 2
  - (33%) RV & unobstructed pulmonary blood flow: 2
  - (33%) LV & obstructed pulmonary blood flow: 3
  - (18%) RV & obstructed pulmonary blood flow: 4
<table>
<thead>
<tr>
<th></th>
<th>Group 1</th>
<th></th>
<th>Group 2</th>
<th></th>
<th>Group 3</th>
<th></th>
<th>Group 4</th>
<th></th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Unobstructed Qp</td>
<td></td>
<td>Unobstructed Qp</td>
<td></td>
<td>Obstructed Qp</td>
<td></td>
<td>Obstructed Qp</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>LV type</td>
<td></td>
<td>RV type</td>
<td></td>
<td>LV type</td>
<td></td>
<td>RV type</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>n = 10</td>
<td></td>
<td>n = 17</td>
<td></td>
<td>n = 23</td>
<td></td>
<td>n = 6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total-fibrosis score</td>
<td>2*</td>
<td></td>
<td>2^</td>
<td></td>
<td>3^</td>
<td></td>
<td>4*^</td>
<td></td>
<td>*0.031</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(0–6)</td>
<td></td>
<td>(0–8)</td>
<td></td>
<td>(2–6)</td>
<td></td>
<td>(1–8)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>*^ Statistical comparisons</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fontan duration, years</td>
<td>15</td>
<td></td>
<td>10</td>
<td></td>
<td>14</td>
<td></td>
<td>17</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(5–21)</td>
<td></td>
<td>(4–24)</td>
<td></td>
<td>(6–29)</td>
<td></td>
<td>(5–22)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Males, %</td>
<td>100</td>
<td></td>
<td>65</td>
<td></td>
<td>43</td>
<td></td>
<td>66</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Oxygen saturation, %</td>
<td>93</td>
<td></td>
<td>92</td>
<td></td>
<td>93</td>
<td></td>
<td>91</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(88–96)</td>
<td></td>
<td>(81–97)</td>
<td></td>
<td>(82–97)</td>
<td></td>
<td>(88–96)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Pacemaker, %</td>
<td>20</td>
<td></td>
<td>18</td>
<td></td>
<td>13</td>
<td></td>
<td>33</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Decreased ventricular fxn, %</td>
<td>30</td>
<td></td>
<td>12</td>
<td></td>
<td>17</td>
<td></td>
<td>33</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>≥Moderate AVVR, %</td>
<td>10</td>
<td></td>
<td>12</td>
<td></td>
<td>0</td>
<td></td>
<td>0</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>IVCP, mmHg</td>
<td>13</td>
<td></td>
<td>13</td>
<td></td>
<td>14</td>
<td></td>
<td>14</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(11–19)</td>
<td></td>
<td>(11–22)</td>
<td></td>
<td>(10–22)</td>
<td></td>
<td>(12–17)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>UVEDP, mmHg</td>
<td>9</td>
<td></td>
<td>8</td>
<td></td>
<td>8</td>
<td></td>
<td>7</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(6–12)</td>
<td></td>
<td>(5–16)</td>
<td></td>
<td>(5–12)</td>
<td></td>
<td>(6–9)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Qs, L/min/m²</td>
<td>2.8</td>
<td></td>
<td>2.6</td>
<td></td>
<td>3.0</td>
<td></td>
<td>2.5</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(1.6–4.5)</td>
<td></td>
<td>(1.9–4.2)</td>
<td></td>
<td>(1.7–4.9)</td>
<td></td>
<td>(1.9–4.2)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>TPP, mmHg</td>
<td>5</td>
<td></td>
<td>5</td>
<td></td>
<td>5</td>
<td></td>
<td>6</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(3–9)</td>
<td></td>
<td>(2–7)</td>
<td></td>
<td>(2–8)</td>
<td></td>
<td>(4–7)</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>PVR, Wood units</td>
<td>1.8</td>
<td></td>
<td>1.7</td>
<td></td>
<td>1.7</td>
<td></td>
<td>1.8</td>
<td></td>
<td>NS</td>
</tr>
<tr>
<td>Median (range)</td>
<td>(0.8–4.5)</td>
<td></td>
<td>(0.9–2.7)</td>
<td></td>
<td>(0.6–4.2)</td>
<td></td>
<td>(1.3–3.5)</td>
<td></td>
<td>NS</td>
</tr>
</tbody>
</table>

AVVR atrioventricular valve regurgitation; fx function; IVCP inferior vena cava pressure; LV type univentricule of left ventricular type; PVR pulmonary vascular resistance; Qp pulmonary blood flow; Qs systemic blood flow; RV type univentricule of right-ventricular type; TPP transpulmonary-pressure gradient; UVEDP univentricular end-diastolic pressure
Sheila Haworth 1977
PLE & Obstructed Pulmonary Blood flow

  - 73% of PLE patients HX of obstructed pulmonary flow

- Our series of Fontans
  - 75% of PLE patients HX of obstructed pulmonary flow
Thank You!

6th Annual Reno Heart Conference

William Evans
Professor Pediatrics University of Nevada