CLINICAL CHARACTERISTICS AND OUTCOME OF HEART TRANSPLANT IN ADULTS WITH CONGENITAL HEART DISEASES

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Heart Transplant for CHD

- Long-term survival in patients with CHD has improved with advances in medical and surgical therapies.
- Currently approximately 85% of children with CHD are surviving to adulthood.
- Progressive cardiopulmonary dysfunction often occurs late after palliative or corrective surgeries and it is the most common cause of death in patients with CHD.
- Consequently, a growing number of patients with complex CHD will require heart transplantation for end-stage heart failure.
Heart Transplant in Adults with CHD

- Objective:
  To analyze outcome in Spanish adult transplanted population with CHD (different subgroups) and compare it with the most frequent causes of adult transplantation:
  - Ischemic Heart Disease [IHD]
  - Idiopathic Dilated Cardiomyopathy [IDCM]
Subanalysis Spanish Registry on Heart Transplantation (1984-2010)

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Heart Transplant in Adults with CHD

Material and Methods

• From May 1984-Dec 2009
  – Total HTx 6048 patients
  – Excluded: Pediatric transplants(<16 years), combined transplants, reHTx, HTx performed in other centers without adult CHD cases, and HTx for heart diseases other than IDCIM and IHD

• Study population: 3166 patients:
  • IHD 1.888; IDCIM 1223: CHD 55
Heart Transplant in Adults with CHD

Material and Methods

Htx 1984-2009
Nº: 6,048

Htx < 16 años
Nº: 239

Heart-Lung Tx: 56
Renal+Cardiac Tx: 46
Liver+Cardiac Tx: 7
ReHtx: 153

HTx without CHD
(excluded)
Nº: 1,301

HTx for other heart diseases
Nº: 1,080

Ischemic Heart Diseases (IHD)
Nº: 1,888

Idiopathic Dilated Cardiomyopathy (IDCM)
Nº: 1,223

Adult Congenital Heart Disease (CHD)
Nº: 55
Heart Transplant in Adults with CHD
Material and Methods

- **Study subgroups:** CHD transplants were classified according to pathophysiology into 4 groups:
  
  1. Single ventricle with different degrees of pulmonary Stenosis (n:18)
  2. Single ventricle with Cavopulmonary shunt surgery (*Glenn/Fontan procedure*) (n:10)
  3. Congenitally corrected transposition of great arteries or DTG with atrial Switch (*Mustard/Senning surgery*) (n:10).
  4. CHDs with different degrees of right ventricle overload *Tetralogy of Fallot, Ebstein’s disease, DTG with PS and Rastelli patch, DORV*) (n:17).
Heart Transplant in Adults with CHD

Results

Significant differences were found between the clinical profile of patients with CHD and other groups: younger age, less need for inotropes and minor renal dysfunction, with less risk factors

<table>
<thead>
<tr>
<th>Clinical profile of patients with Adult Heart Transplantation</th>
<th>IHD</th>
<th>IDCm</th>
<th>CHD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)#</td>
<td>56 (11)</td>
<td>54 (16)</td>
<td>26 (25)</td>
</tr>
<tr>
<td>Male#</td>
<td>91</td>
<td>79</td>
<td>62</td>
</tr>
<tr>
<td>Weight (Kg)#</td>
<td>73 (15)</td>
<td>72 (17)</td>
<td>59 (19)</td>
</tr>
<tr>
<td>Inotropes e.v.*</td>
<td>34</td>
<td>32</td>
<td>16</td>
</tr>
<tr>
<td>Renal Dysfunction (Cr&gt;2)*</td>
<td>13</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary Hypertension</td>
<td>47 (22)</td>
<td>46 (17)</td>
<td>35 (21)</td>
</tr>
<tr>
<td>Bilirrub. (&gt;2.5 mg/dL)#</td>
<td>13</td>
<td>18</td>
<td>32</td>
</tr>
<tr>
<td>ALT/ AST (&gt;100 mg/dL)</td>
<td>24</td>
<td>27</td>
<td>21</td>
</tr>
<tr>
<td>Diabetes#</td>
<td>16</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>Arterial Hypertension#</td>
<td>38</td>
<td>21</td>
<td>7</td>
</tr>
<tr>
<td>Dyslipidemia#</td>
<td>52</td>
<td>26</td>
<td>11</td>
</tr>
<tr>
<td>Ventricular Asistance#</td>
<td>17</td>
<td>11</td>
<td>9</td>
</tr>
</tbody>
</table>

* p<0.05
# p<0.001
(Spanish Registry on Heart Transplantation: 1984-2010)
Heart Transplant in Adults with CHD

Results

- The analysis of survival showed some differences among groups (CHD vs IHD, p=0.05; CHD vs IDCMM, p=0.5; IHD vs IDCMM, p=0.0001)
- The probability of overall survival was higher in CHD group, despite its high early mortality.
Heart Transplant in Adults with CHD

Results

- Early mortality was different between the different CHD subgroups (group 1: 19%, group 2: 40%, group 3: 0%, group 4: 29%; p<0.001)
- No significant differences among subgroups were found in long-term survival

Group 1: Single ventricle + PS
Group 2: SV + Glenn/ Fontan surgery
Group 3: RV systemic (CTGA/ Senning)
Group 4: CHDs overload RV
Heart Transplant in Adults with CHD

Summary

• Percentage of adult with CHD transplanted in Spain is low (1%).

• Long term survival is higher compared with groups with different indications for HTx (IHD and IDCM).

• Early mortality is high in operated patients with single ventricle physiology.

• European registries with a larger number of patients are needed to better define results in this group of patients.
Thank you for your attention!

Vielen Dank!  Grazie Mille!

Muchas Gracias!