SURVIVAL AND REVERSE CARDIAC REMODELLING IN CHILDHOOD DILATED CARDIOMYOPATHY (DCM): RESULTS FROM A LONG-TERM NATIONAL POPULATION-BASED STUDY

ACC Moderated Poster Contributions
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Background: DCM in childhood is characterised by varied causes and heterogeneous outcomes. The incidence of normalisation of cardiac function over time has not been well defined.

Methods: The National Australian Childhood Cardiomyopathy Study is a longitudinal national study of all children diagnosed with primary cardiomyopathy at age 0-10 years, in 1987-1997. Serial echo parameters of LV size and fractional shortening were recorded. Normalization of LV function was defined as LVED Z score -2.0. Competing risk analysis was undertaken for the 3 mutually exclusive states: alive with normalisation of LV function, alive without normalisation, and death/ transplantation. All cardiac histology collected during life was reviewed centrally.

Results: There were 184 patients with DCM. Of these, 9 (5%) presented with sudden death and were not included in this analysis. The median duration of follow-up was 15.1 years (IQR 13.0, 17.3) years.

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<tr>
<th></th>
<th>n</th>
<th>Alive, normalisation</th>
<th>Alive, no normalisation</th>
<th>Death/ transplant</th>
</tr>
</thead>
<tbody>
<tr>
<td>All DCM patients</td>
<td>175</td>
<td>30%</td>
<td>35%</td>
<td>35%</td>
</tr>
<tr>
<td>Myocarditis positive</td>
<td>13</td>
<td>90%</td>
<td>10%</td>
<td>0%</td>
</tr>
<tr>
<td>Myocarditis negative</td>
<td>27</td>
<td>30%</td>
<td>34%</td>
<td>36%</td>
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</tbody>
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Conclusions: At long-term follow-up, 30% of children with DCM have normal left ventricular systolic function. Survival and reverse remodelling are better in those with biopsy proven myocarditis.