Sigma’s 30th International Nursing Research Congress
Outcomes of Heart Transplantation and Combined Heart-Liver Transplantation in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy
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Introduction: Existing literature on orthotopic heart transplantation (OHT) in arrhythmogenic right ventricular cardiomyopathy (ARVC) is limited to case reports, a single-center series, and a registry retrospective cohort study.

Purpose: We aimed to define the characteristics and outcomes of ARVC patients who underwent OHT and combined heart-liver transplantation (OHT-OLT) at a U.S. tertiary academic medical center.

Methods: We performed a retrospective analysis of 14 patients in the Penn ARVC Transplant Database. Thirteen patients underwent OHT or OHT-OLT between May 2011 and August 2017. Demographics, pre-transplant characteristics, and survival data were collected. Normality was not assumed in the continuous data fields given the small sample size. Descriptive statistics were computed as medians and interquartile ranges (IQRs). The Wilcoxon rank-sum test and Fisher’s exact test were used for inferential statistical testing of continuous and categorical variables, respectively.

Results: Features of the study population are presented in Table 1. Nine of 13 patients underwent OHT and 4 underwent OHT-OLT. One patient died awaiting transplant. The median ages at diagnosis and listing for transplant were similar. Although not statistically significant, there were trends toward longer diagnosis to listing (127.3 months vs. 48.5 months) and diagnosis to transplant times (130.9 months vs. 51.4 months) in the OHT compared to OHT-OLT group. The post-transplant survival was 100% for both OHT and OHT-OLT with median post-transplant follow-up times of 4.1 years and 2.1 years, respectively.

Table 1: Characteristics and Outcomes by Transplant Type

<table>
<thead>
<tr>
<th>Variables</th>
<th>Heart</th>
<th>Heart-Liver</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>9</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Age at Diagnosis (years), median (IQR)</td>
<td>50.0 (21.0, 55.5)</td>
<td>49.5 (34.5, 55.0)</td>
<td>0.88</td>
</tr>
<tr>
<td>Age at Listing (years), median (IQR)</td>
<td>59.0 (55.0, 61.0)</td>
<td>55.9 (38.0, 59.5)</td>
<td>0.44</td>
</tr>
<tr>
<td></td>
<td>Female</td>
<td>Male</td>
<td>p-value</td>
</tr>
<tr>
<td>---------------------------</td>
<td>--------</td>
<td>--------</td>
<td>---------</td>
</tr>
<tr>
<td>Sex</td>
<td>3 (33%)</td>
<td>6 (67%)</td>
<td>1.00</td>
</tr>
<tr>
<td>Female</td>
<td>1 (25%)</td>
<td>3 (75%)</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transplant Status</td>
<td></td>
<td></td>
<td>0.41</td>
</tr>
<tr>
<td>1A</td>
<td>0 (0%)</td>
<td>1 (25%)</td>
<td></td>
</tr>
<tr>
<td>1B</td>
<td>3 (33%)</td>
<td>1 (25%)</td>
<td></td>
</tr>
<tr>
<td>1B by exception</td>
<td>5 (56%)</td>
<td>1 (25%)</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>1 (11%)</td>
<td>1 (25%)</td>
<td></td>
</tr>
<tr>
<td>BMI (kg/m²), median (IQR)</td>
<td>23.4 (20.6, 28.8)</td>
<td>27.5 (26.9, 27.7)</td>
<td>0.26</td>
</tr>
<tr>
<td>Creatinine (mg/dL), median (IQR)</td>
<td>1.1 (1.0, 1.3)</td>
<td>1.2 (1.2, 2.0)</td>
<td>0.31</td>
</tr>
<tr>
<td>MELD, median (IQR)</td>
<td>13.0 (12.0, 15.0)</td>
<td>18.5 (13.5, 19.0)</td>
<td>0.33</td>
</tr>
<tr>
<td>SVR (dyn-s/cm²), median (IQR)</td>
<td>2014.5 (1222.5, 2864.0)</td>
<td>1264.0 (1255.0, 1760)</td>
<td>0.31</td>
</tr>
<tr>
<td>Diagnosis to Listing (months), median (IQR)</td>
<td>127.3 (49.1, 153.5)</td>
<td>48.5 (27.7, 77.8)</td>
<td>0.12</td>
</tr>
<tr>
<td>Diagnosis to Transplant (months), median (IQR)</td>
<td>130.9 (56.6, 154.8)</td>
<td>51.4 (40.6, 58.3)</td>
<td>0.17</td>
</tr>
<tr>
<td>Time on Wait List (days), median (IQR)</td>
<td>110.0 (59.0, 154.0)</td>
<td>74.5 (27.5, 588.5)</td>
<td>0.76</td>
</tr>
<tr>
<td>Survival?</td>
<td></td>
<td></td>
<td>0.31</td>
</tr>
<tr>
<td>No</td>
<td>0 (0%)</td>
<td>1 (25%)</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>9 (100%)</td>
<td>3 (75%)</td>
<td></td>
</tr>
</tbody>
</table>
Conclusion: ARVC can be managed by collaboration of MD and APP. The basic understanding of genetic predispositions and risks factors will aid in early detection, early referral and prompt treatment of ARVC by an expert cardiologist for early referral for possible heart alone or combine heart-liver transplantation.

Title:
Outcomes of Heart Transplantation and Combined Heart-Liver Transplantation in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy

Keywords:
Arrhythmogenic right ventricular cardiomyopathy, Combined heart-liver transplant and heart transplant

References:


Abstract Summary:
Existing literature on orthotopic heart transplantation (OHT) in arrhythmogenic right ventricular cardiomyopathy (ARVC) is limited to case reports, a single center series, and a registry retrospective cohort study. This presentation will define the characteristics and outcomes of ARVC patients who underwent OHT and combined heart-liver transplantation.
Presentation Title: Outcomes of Heart Transplantation and Combined Heart-Liver Transplantation in Patients with Arrhythmogenic Right Ventricular Cardiomyopathy

I. Introduction
   a. Background and Significance
      1. Literature on orthotopic heart transplantation (OHT) in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC) is limited to case reports, single-center series, and a registry retrospective cohort study.
      2. Advance Practice Nurses are having increasing roles in the collaborative care of patients with ARVC requiring heart transplantation.
   b. Review of Basics of ARVC
      1. Definition and Pathophysiology of ARVC
      2. Prevalence of ARVC
      3. MELD score Stratification of heart vs combined heart-liver transplant

II. Study Design and Methods
   a. Retrospective Analysis of 14 patients in the Penn ARVC Transplant Database
   b. Demographics, pre-transplant characteristics and survival data were collected.
   c. Descriptive statistics were computed as medians and interquartiles ranges. The Wilcoxon rank-sum test and fisher exact test were used for inferential statistical testing.

IV. Results
   a. 9 of 13 patients underwent OHT and 4 underwent OHT-OLT, 1 died awaiting transplant
   b. Median ages at diagnosis and listing for transplant were similar
   c. The post transplant survival was 100% for both OHT and OHT-OLT with median post transplant follow-up times of 4.1 years and 2.1 years respectively.
   d. Discussion of Table on Characteristics and Outcomes by Transplant Type

V. Conclusion
   a. ARVC can be managed by collaboration with physicians and advance practice nurses.
   b. The basic understanding of genetic predispositions and risks factors will aid in early detection, early referral and prompt treatment of ARVC by an expert cardiologist for early referral for possible heart alone of combined heart-liver transplantation.

VI Implications for Practice
   a. Advance Practice Nurse role delineation and collaborative practices
   b. Team based approach for ARVC management: cardiac surgery, critical care, advanced heart failure, hepatology and allied health practitioners

First Primary Presenting Author

Primary Presenting Author

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**Author Summary:** Maria Molina has been an acute care nurse practitioner in the field of advanced heart disease specifically heart and lung transplant for the past decade. She is an active nurse researcher and is well published in the field of heart, lung transplant and immunology. She has also been the recipient of awards recognizing her clinical expertise and passion for research.

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**Author Summary:** Ylenia Quiaoit has been practicing as an acute care nurse practitioner in the field of advanced heart failure and transplant for the last 15 years, and has been serving as clinical faculty with University of Pennsylvania acute care NP program. She has presented nationally and internationally on topics related to advanced HF, transplant and palliative care.

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**Author Summary:** She has been a nurse practitioner for over a decade in the area of heart transplantation. She is currently the supervisor of the heart transplant advance practice team at the Hospital of the University of Pennsylvania. She has presented nationally and internationally on topics related the heart transplantation and advanced heart disease.