**Supplementary Table 1:** Sociodemographic, clinical profile, symptoms, and MRI assessment in our study population. Comparison between alive and succumbed patients.

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Parameter** | **All-Population**  **(n=57)** | **Alive**  **(n=45)** | **Succumbed**  **(n=12)** | ***P*-value** |
| Men (%) | 30 (52.63) | 23 (51.11) | 7 (58.33) | 0.904 |
| Age (Years) | 29.52 (21.24-39.17) | 30.15 (20.79-39.17) | 27.26 (23.54-40.36) | 0.462 |
| Age at Diagnosis (Years) | 18.83 (13.32-30.39) | 18.62 (10.48-25.7) | 23.54 (17.74-33.7) | 0.144 |
| Time of Treatment (Years) | 9.32 (4.67-12.98) | 10.16 (4.91-13.61) | 5.2 (4.43-8.93) | 0.094 |
| Clinical Profile | | | | |
| BSA (m2/1.73) | 1.68 (± 0.2) | 1.7 (± 0.19) | 1.63 (± 0.24) | 0.213 |
| BMI (Kg/m2) | 24.1 (± 4.12) | 24.17 (± 4.24) | 23.87 (± 3.8) | 0.976 |
| Underweight (%) | 4 (7.02) | 3 (6.67) | 1 (8.33) | 0.912 |
| Normal weight (%) | 34 (59.65) | 26 (57.78) | 8 (66.67) | 0.812 |
| Overweight (%) | 13 (22.81) | 11 (24.44) | 2 (16.67) | 0.984 |
| Obesity (%) | 6 (10.53) | 5 (11.11) | 1 (8.33) | 0.764 |
| Diabetes (%) | 1 (1.75) | 1 (2.22) | 0 (0) | 0.999 |
| Arterial Hypertension (%) | 4 (7.02) | 4 (8.89) | 0 (0) | 0.568 |
| Stroke (%) | 4 (7.02) | 4 (8.89) | 0 (0) | 0.568 |
| Other Comorbidity (%) | 17 (29.82) | 12 (26.67) | 5 (41.67) | 0.478 |
| Wolff-Parkinson White (%) | 17 (29.82) | 16 (35.56) | 1 (8.33) | 0.085 |
| Atrio-Ventricular Block (%) | 7 (12.28) | 6 (13.33) | 1 (8.33) | 0.981 |
| Atrial Fibrillation (%) | 8 (14.04) | 4 (8.89) | 4 (33.33) | 0.052 |
| Right Bundle Branch Block (%) | 4 (7.02) | 3 (6.67) | 1 (8.33) | 0.731 |
| Other Arrhythmia (%) | 4 (7.02) | 3 (6.67) | 1 (8.33) | 0.683 |
| Interatrial-Communication (%) | 32 (56.14) | 24 (53.33) | 8 (66.67) | 0.520 |
| Interventricular-Communication (%) | 1 (1.75) | 1 (2.22) | 0 (0) | 0.951 |
| Patent Foramen Ovale (%) | 13 (22.81) | 11 (24.44) | 2 (16.67) | 0.713 |
| Other Structural Impairment (%) | 17 (29.82) | 12 (26.67) | 5 (41.67) | 0.941 |
| Symptoms | | | | |
| Dyspnea (%) | 13 (22.81) | 7 (15.56) | 6 (50) | 0.020 |
| Palpitations (%) | 18 (31.58) | 13 (28.89) | 5 (41.67) | 0.489 |
| Edema (%) | 5 (8.77) | 2 (4.44) | 3 (25) | 0.057 |
| Fatigue (%) | 10 (17.54) | 7 (15.56) | 3 (25) | 0.424 |
| Tachycardia (%) | 2 (3.51) | 1 (2.22) | 1 (8.33) | 0.379 |
| Cyanosis (%) | 9 (15.79) | 6 (13.33) | 3 (25) | 0.379 |
| NYHA I (%) | 46 (80.7) | 40 (88.89) | 6 (50) | 0.006 |
| NYHA II (%) | 10 (17.54) | 5 (11.11) | 5 (41.67) | 0.025 |
| NYHA III (%) | 1 (1.75) | 0 (0) | 1 (8.33) | 0.211 |
| Hemoglobin (gr/dl) | 15.6 (14.3-17.75) | 15.5 (14.25-17.15) | 15.7 (14.6-19.75) | 0.471 |
| Hematocrit (%) | 46.4 (42.25-53.45) | 45.95 (41.53-51.83) | 47 (44-60.95) | 0.436 |
| CMR assessment | | | | |
| Severity Index | 0.7 (0.53-0.95) | 0.64 (0.53-0.95) | 0.88 (0.63-1.46) | 0.243 |
| Septal Leaflet Displacement (mm) | 62 (47-72) | 60 (47-74) | 66 (46.75-68.5) | 0.968 |
| Posterior Leaflet Displacement (mm) | 73 (62-78) | 73 (62-78) | 74 (62-74.75) | 0.961 |
| Right Ventricle - Atrialization (mm) | 63 (50-83) | 53 (50-70) | 83 (69.25-86.25) | 0.017 |
| Right Ventricle - Atrialization (%) | 63 (58-76) | 63 (58-73) | 70.5 (57-76.5) | 0.346 |
| LVEF (%) | 52 (43-57) | 54 (47-59) | 35.5 (24-46.75) | <0.001 |
| RVEF (%) | 34 (25-43) | 36 (27-45) | 25 (13.3-34.75) | 0.021 |
| LV-EDV Indexed (mL/m2) | 51 (41-63) | 51 (40-62.3) | 52.5 (44.75-85.5) | 0.505 |
| LV-ESV Indexed (mL/m2) | 26 (20-31) | 23 (20-30) | 32 (26-47) | 0.007 |
| LV-Mass Indexed (g/m2) | 35 (29-42) | 34 (28-42) | 36 (33.75-67.75) | 0.076 |
| RV-EDV Indexed (mL/m2) | 161 (115-229) | 161 (126-224) | 146.5 (93-297) | 0.914 |
| RV-ESV Indexed (mL/m2) | 120 (79-163) | 120 (64-163) | 148 (113.5-266) | 0.082 |
| Enhanced Areas (n) | 0 (0-1) | 0 (0-0) | 1 (0-2) | 0.031 |
| Left-Atrium (%) | 3 (5.26) | 3 (6.67) | 0 (0) | 0.848 |
| Right-Atrium (%) | 14 (24.56) | 10 (22.22) | 4 (33.33) | 0.463 |
| Left-Ventricle (%) | 17 (29.82) | 10 (22.22) | 7 (58.33) | 0.029 |
| Right-Ventricle (%) | 8 (14.04) | 4 (8.89) | 4 (33.33) | 0.052 |
| Intra-myocardial (%) | 14 (24.56) | 8 (17.78) | 6 (50) | 0.053 |
| Subendocardial (%) | 1 (1.75) | 0 (0) | 1 (8.33) | 0.211 |
| Subepicardial (%) | 1 (1.75) | 1 (2.22) | 0 (0) | 0.912 |
| Transmural (%) | 4 (7.02) | 3 (6.67) | 1 (8.33) | 0.124 |

BMI, body mass index; BSA, body surface area; CMR, cardiac magnetic resonance; EDV, end-diastolic volume; ESV, end-systolic volume; LV, left ventricle; LVEF, left ventricle ejection fraction; NYHA, New York Heart Association; RV, right ventricle; RVEF, right ventricle ejection fraction.

**Supplementary Table 2:** Cox proportional hazard regression models to LV-LGE as a predictor of mortality in patients with Ebstein’s Abnormality.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Model Parameters** | **Predictor** | **Coefficients** | **HR** | **95% CI** | **P Value** |
| **Unadjusted**  R2=0.082  Dxy= 0.213  C-Statistic: 0.606  BIC: 69.08207  Wald Test (P-Value): <0.001 | LV-Fibrosis | 1.133 | 3.106 | 0.90-10.68 | 0.072 |
| **Model 1**  R2=0.281  Dxy= 0.599  C-Statistic: 0.884  BIC: 57.53123  Wald Test (P-Value): <0.001 | LV-Fibrosis | 1.431 | 4.181 | 1.14-15.30 | 0.030 |
| Male Sex | 2.676 | 14.539 | 2.02-104.1 | <0.001 |
| BSA | -3.810 | 0.022 | 0.003-0.993 | 0.042 |
| **Model 2**  R2=0.317  Dxy= 0.605  C-Statistic: 0.888  BIC: 62.93864  Wald Test (P-Value): <0.001 | LV-Fibrosis | 1.593 | 6.020 | 1.22-19.91 | 0.025 |
| Male Sex | 2.718 | 15.155 | 1.65-138.5 | 0.016 |
| BSA | -4.821 | 0.008 | 0.001-0.609 | 0.028 |
| Number of comorbidities | -0.147 | 0.863 | 0.389-1.91 | 0.717 |
| LVEF | -0.078 | 0.9246 | 0.881-0.969 | 0.001 |
| LV-Mass | -0.022 | 0.977 | 0.941-1.016 | 0.256 |
| RV-Atrialization | -0.027 | 0.972 | 0.918-1.030 | 0.344 |
| Time since arrival at the institute | -0.191 | 0.825 | 0.682-1.002 | 0.084 |

**Supplementary Table 3:** Cox proportional hazard regression models to assess the risk for mortality associated with the mortality score for Ebstein’s Abnormality.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
| **Model Parameters** | **Predictor** | **Coefficients** | **HR** | **95% CI** | **P Value** |
| **Individual components**  R2=0.436  Dxy= 0.854  C-Statistic: 0.927  BIC: 58.93449  Wald Test (P-Value): <0.001 | Male Sex (Yes) | 2.865 | 17.55 | 2.456-125.4 | 0.004 |
| BSA (𝑚2/1.73) | -4.326 | 0.013 | 0.01-0.988 | 0.048 |
| RVEF (%) | -0.068 | 0.934 | 0.889-0.981 | 0.006 |
| LV Fibrosis (Yes) | 1.292 | 3.642 | 1.021-13.00 | 0.046 |
| **Continuous**  R2=0.435  Dxy= 0.860  C-Statistic: 0.93  BIC: 51.51224  Wald Test (P-Value): <0.001 | Mortality Score Equation | 0.6867 | 1.987 | 1.425-2.771 | <0.001 |
| **Categorical Variable**  R2=0.251  Dxy= 0.514  C-Statistic: 0.757  BIC: 61.31954  Wald Test (P-Value): <0.001 | Mortality Score ≥6.5 | 2.611 | 13.62 | 1.715-108.1 | 0.013 |

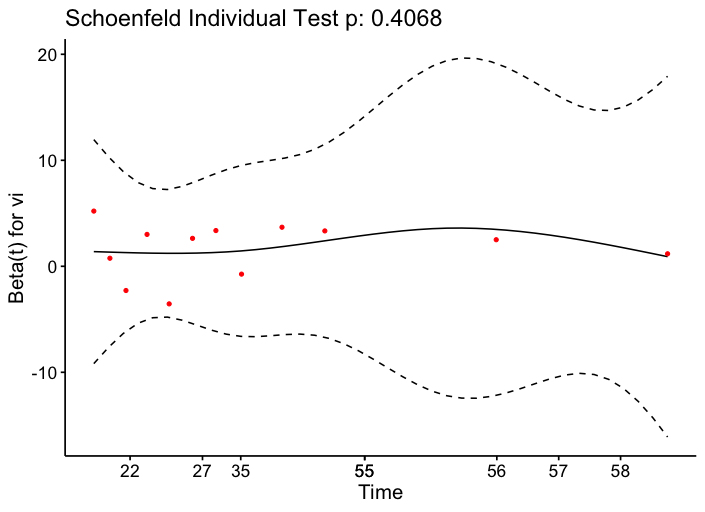
.

**Supplementary Figure 1: Schoenfeld residuals to test the proportional hazard assumption parameter from the** Cox proportional hazard regression model’s metrics assess the risk for mortality associated with LV-LGE (+). *Model 1*: adjusted for sex and body surface area. *Model 2*: adjusted for model 1 + number of commodities, LVEF, LV Mass Index and RV atrialization.

Chart, line chart

Description automatically generatedChart, line chart

Description automatically generated



Abbreviations: BIC= Bayesian Information Criteria; SE= Standard Error.

**Supplementary Figure 2:** Kaplan-Meier analysis to evaluate the survival probability associated with a mortality score <6.5 pts.

