**Supplemental Figures**

Supplemental Fig1

Fig. 1 Patients included in the study by their diagnosis, enumerating the patients within each diagnosis. Miscellaneous 2V patients included diagnoses such as pulmonic valve, artery, or vein stenosis, mitral stenosis, and double outlet right ventricle

Supplemental Fig2

Fig2. The percentage of individuals with an abnormal FVC based on whether they have had at most two or more than three cardiac surgeries and whether they have single or two ventricle circulation

**Supplemental Tables**

Supplemental Table 1. Patient demographics of single and two ventricle populations.

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | **Total****n=260** | **Single Ventricle****n=80** | **Two Ventricle****n=180** | ***p* Value** |
| Age in years at testing, median (IQR) | 14.2 (12.0-17.2) | 13.6 (11.5-16.8) | 14.4 (12.0-17.3) | 0.243 |
| Sex, n (%) |  |  |  |  |
| Male | 166 (65.4%) | 52 (65.0%) | 118 (65.6%) | 0.931 |
| Female | 88 (34.6%) | 28 (35.0%) | 62 (34.4%) |  |
| BMI, median (IQR) |  |  |  |  |
| z-score\* | 0.33 (-0.55-1.17) | 0.28 (-0.54-1.31) | 0.36 (-0.58-1.12) | 0.972 |
| Ethnicity, n (%) |  |  |  |  |
| Hispanic | 132 (53.1%) | 44 (61.3%) | 88 (49.4%) | 0.078 |
| Non-Hispanic | 122 (46.9%) | 31 (38.8%) | 91 (50.6%) |  |

\* BMI Z-score data not reported in 13 patients due to age

Supplemental Table 2. Demographic data of Single Ventricle, Tetralogy of Fallot and Truncus Arteriosus populations

|  |  |  |  |
| --- | --- | --- | --- |
|  | **Single Ventricle** | **Tetralogy of Fallot** | **Truncus Arteriosus** |
| n (% of sample) | 80 (30.8%) | 48 (35.3%) | 8 (3.1%) |
| Age at Testing (years), median (IQR) | 13.6 (11.5-16.8) | 14.2 (11.6-17.8) | 13.7 (11.4-18.0) |
| Sex, n (%) |  |  |  |
| Male | 52 (65.0%) | 34 (70.8%) | 4 (50%) |
| Female | 28 (35.0%) | 14 (29.2%) | 4 (50%) |
| BMI, median (IQR) |  |  |  |
| z-scorea | 0.28 (-0.54-1.31) | -0.18 (-0.86-0.88) | -0.65 (-1.6-0) |
| Ethnicity, n (%) |  |  |  |
| Hispanic | 49 (61.3%) | 18 (37.5%) | 4 (50%) |
| Non-Hispanic | 31 (38.8%) | 30 (62.5%) | 4 (50%) |

Supplemental Table 3. Demographic data of patients with abnormal and normal FVC

|  |  |  |  |
| --- | --- | --- | --- |
|  | **Abnormal FVC****n=86** | **Normal FVC****n=174** | ***p* Value** |
| Age at Testing (years), median (IQR) | 14.4 (12.0-17.7) | 14.0 (11.9-16.7) | 0.178 |
| Sex, n (%) |  |  |  |
| Male | 53 (61.6%) | 117 (67.2%) | 0.371 |
| Female | 33 (38.4%) | 57 (32.8%) |  |
| BMI, median (IQR) |  |  |  |
| z-score | 0.48 (-0.23-1.31) | -0.52 (-1.3-0.89) | <0.001 |
| Ethnicity, n (%) |  |  |  |
| Hispanic | 44 (51.2%) | 94 (54.0%) | 0.664 |
| Non-Hispanic | 42 (48.8%) | 80 (46.0%) |  |

Supplemental Table 4. Demographics of subjects by age

|  |  |  |  |
| --- | --- | --- | --- |
|  | **Age < 18 years old** | **Age ≥ 18** | ***p* Value** |
| n (%) | 218 (83.9%) | 42 (16.2%) |  |
| Age at Testing (years) | 13.2 (11.5-15.6) | 19.7 (18.7-20.5) | <0.001 |
| Sex, n (%) |  |  |  |
| Male | 144 (66.1%) | 26 (61.9%) | 0.605 |
| Female | 74 (33.9%) | 16 (38.1%) |  |
| BMI, median (IQR) |  |  |  |
| z-scorea | 0.25 (-0.62-1.10) | 0.95 (-0.10-1.57) | 0.007 |
| Ethnicity, n (%) |  |  |  |
| Hispanic | 112 (51.4%) | 26 (61.9%) | 0.211 |
| Non-Hispanic | 106 (48.6%) | 16 (38.1%) |  |
| FVC z-score, median (IQR) | -1.02 (-1.90- -0.22) | -1.49 (-2.35 - -0.49) | 0.078 |
| Abnormal z-score FVC, n (%)  | 67 (30.7%) | 19 (45.2%) | 0.067 |
|  FEV1sec /FVC z-score, median (IQR) | 0.27 (-0.57-1.29) | 0.081 (-0.36-0.73) | 0.330 |
| Abnormal z-score FEV1sec /FVC, n (%) | 10 (4.6%) | 1 (2.38%) | 0.515 |
| Cardiac surgeries, median (IQR) | 2 (1-3) | 3 (1-3) | 0.1165 |

Supplemental Table 5. Spirometry and surgical data of TOF, TA, and all other 2V diagnoses of children under the age of 18.

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | **Single Ventricle****n=68** | **Tetralogy of Fallotn=37** | **Truncus Arteriosusn=6** | **Other 2V****n=107** |
| FVC % pred, median (IQR) | 83.5 (74.8-92.9) | 82.1 (71.0-92.7)p = 0.5279 | 79.5 (66.4-81.5)p = 0.1606 | 93.7 (83.2-100.4)p < 0.001 |
| FVC z-score, median (IQR) | -1.4 (-2.2- -0.6) | -1.57 (-2.54- -0.63)p = 0.4736 | -1.74 (-2.94- -1.62)p = 0.1168 | -0.55 (-1.5-0.03)p < 0.001 |
| Abnormal z-score FVC, n (%) | 28 (41.2%) | 16 (43.2%)p=0.838 | 4 (66.7%)p = 0.227 | 19 (17.8%)p = 0.001 |
| FEV1sec % pred, median (IQR) | 85.8 (77.7-93.8) | 86.5 (74.6-95.6)p = 0.5367 | 72.5 (61.1-83.5)p = 0.0573 | 93.87 (84.5-101.8)p = 0.009 |
| FEV1sec /FVC, median (IQR) | 90.5 (86-93.5) | 90 (83-94)p = 0.6194 | 82.5 (80-84)p = 0.0182 | 88 (83-94)p = 0.0376 |
| Abnormal z-score FEV1sec /FVC, n (%) | 1 (1.5%) | 2 (5.4%)p = 0.248 | 1 (16.7%)p = 0.028 | 6 (5.6%)p = 0.173 |
| FEF25-75% pred, median (IQR) | 92.3 (76.7-104.7) | 89.6 (70.2-107.3)p = 0.6048 | 59.4 (51.7-66.2)p = 0.0171 | 96.1 (78.4-111.4)p = 0.4570 |
| Cardiac surgeries, median (IQR) | 3 (3-4) | 2 (1-2)p<0.001 | 2.5 (1-3)p = 0.0108 | 1 (0-1)p<0.001 |

p Values are calculated from single ventricle data against either TOF, TA, or Other 2V.