**Appendix 1: Supplementary Materials**

Table S1. Results from univariable logistic regression analyses of candidate variables:

|  | **Goodness-of-fit**  **measures** | |  | **Leave-one-out**  **cross validation** | |  | **Measures of discriminative ability (95%CI)** | | |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Echocardiogram**  **parameter** | **R2** | **Residual**  **Deviance** | **AIC** | **Brier score** | **BSS** |  | **AUC** | **Sensitivitya** | **Specificityb** |
| RV/LV length ratio | 0.50 | 30.4 | 34.4 | 0.13 | 0.45 |  | 0.91  (0.80-1.00) | 73.3%  (48.0-89.1%) | 92.0%  (75.0-97.8%) |
| RV cavity lengthc | 0.43 | 33.1 | 37.1 | 0.15 | 0.36 |  | 0.89  (0.77-1.00) | 73.3%  (48.0-89.1%) | 88.0%  (70.0-95.8%) |
| TV annulus mm | 0.42 | 32.3 | 36.3 | 0.15 | 0.35 |  | 0.88  (0.76-1.00) | 66.7%  (41.7-84.8%) | 84.0%  (65.3-93.6%) |
| TV effective orifice | 0.41 | 34.2 | 38.2 | 0.16 | 0.34 |  | 0.82  (0.68-0.97) | 66.7%  (41.7-84.8%) | 96.0%  (80.5-99.3%) |
| TV/MV annulus ratio | 0.40 | 33.0 | 37.0 | 0.16 | 0.33 |  | 0.88  (0.76-1.00) | 60.0%  (35.7-80.2%) | 84.0%  (65.3-93.6%) |
| TV annulus z-score | 0.39 | 34.3 | 38.3 | 0.16 | 0.32 |  | 0.88  (0.75-1.00) | 66.7%  (41.7-84.8%) | 84.0%  (65.3-93.6%) |
| MPA diameter | 0.24 | 41.9 | 45.9 | 0.20 | 0.17 |  | 0.79  (0.63-0.94) | 60.0%  (35.7-80.2%) | 76.0%  (56.6-88.5%) |
| MV annulus mm | 0.00 | 52.9 | 56.9 | 0.26 | -0.10 |  | 0.54  (0.35-0.73) | 37.5%  (24.2-53.0%) | NA%  (NA-NA%) |
| aThe proportion of biventricular repairs correctly predicted; bThe proportion of univentricular repairs correctly predicted; cLength of cavity minus trabeculations.  R2: Efron’s pseudo-R2; AIC: Akaike Information Criterion; BBS: Brier skill score; AUC: area under the receiver operating curve; RV: right ventricle; LV: left ventricle; TV: tricuspid valve; MV: mitral valve; MPA: main pulmonary artery; PV: pulmonary valve. | | | | | | | | | |

Compared to univariable results (Table S1), improved model and discriminative performance was seen for nine of the candidate models considered in bivariable logistic regression modelling, with significant independent contributions to prediction from each in the respective pairs compared to either alone (Table S2).

The highest value of Efron’s pseudo- was the criterion used for selection of the final model out of the candidate models considered, yielding a discriminatory score based on the ratio of the right to left ventricle length and the ratio of the tricuspid to mitral valve dimensions (Table S2):

Table S2. Results from bivariable logistic regression analyses in which each of the pair significantly contributed to the drop in deviance of the joint model compared to the null.

|  | **Goodness-of-fit**  **measures** | |  | **Leave-one-out**  **cross validation** | |  | **Measures of discriminative ability (95%CI)** | | |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Model-derived score** | **R2** | **Deviance** | **AIC** | **Brier** | **BSS** |  | **AUC** | **Sensitivitya** | **Specificityb** |
| 8.5\*RV/LV length ratio +  6.7\*TV/MV annulus ratio - 11.3 | 0.70 | 21.1 | 27.1 | 0.09 | 0.62 |  | 0.95  (0.87-1.00) | 93.3%  (70.2-98.8%) | 96.0%  (80.5-99.3%) |
| 7.6\*RV/LV length ratio +  0.5\*TV annulus mm - 10.2 | 0.66 | 22.9 | 28.9 | 0.10 | 0.58 |  | 0.94  (0.85-1.00) | 86.7%  (62.1-96.3%) | 96.0%  (80.5-99.3%) |
| 8.3\*RV/LV length ratio +  0.7\*TV annulus z-score - 5.2 | 0.66 | 23.0 | 29.0 | 0.10 | 0.58 |  | 0.94  (0.85-1.00) | 86.7%  (62.1-96.3%) | 92.0%  (75.0-97.8%) |
| 6.4\*TV/MV annulus ratio +  0.3\*RV length - 10.3 | 0.63 | 23.8 | 29.8 | 0.11 | 0.54 |  | 0.95  (0.87-1.00) | 80.0%  (54.8-93.0%) | 92.0%  (75.0-97.8%) |
| 0.3\*RV length +  0.7\*TV annulus z-score - 4.6 | 0.61 | 25.1 | 31.1 | 0.11 | 0.51 |  | 0.94  (0.85-1.00) | 80.0%  (54.8-93.0%) | 88.0%  (70.0-95.8%) |
| 0.2\*RV length +  0.5\*TV annulus mm - 9.5 | 0.59 | 25.6 | 31.6 | 0.12 | 0.50 |  | 0.93  (0.83-1.00) | 80.0%  (54.8-93.0%) | 92.0%  (75.0-97.8%) |
| 0.8\*TV annulus z-score +  2.5\*TV effective orifice - 5.7 | 0.58 | 24.4 | 30.4 | 0.13 | 0.46 |  | 0.94  (0.85-1.00) | 80.0%  (54.8-93.0%) | 96.0%  (80.5-99.3%) |
| 7.5\*RV/LV length ratio +  1.6\*TV effective orifice - 8.9 | 0.58 | 24.3 | 30.3 | 0.13 | 0.46 |  | 0.95  (0.87-1.00) | 80.0%  (54.8-93.0%) | 92.0%  (75.0-97.8%) |
| 0.6\*TV annulus mm +  2\*TV effective orifice - 11.2 | 0.57 | 25.0 | 31.0 | 0.13 | 0.45 |  | 0.95  (0.87-1.00) | 86.7%  (62.1-96.3%) | 88.0%  (70.0-95.8%) |
| 6\*TV/MV annulus ratio +  2\*TV effective orifice - 9.9 | 0.54 | 25.4 | 31.4 | 0.14 | 0.42 |  | 0.93  (0.84-1.00) | 73.3%  (48.0-89.1%) | 88.0%  (70.0-95.8%) |
| 2.2\*TV effective orifice +  0.6\*MPA - 10.5 | 0.53 | 27.6 | 33.6 | 0.14 | 0.39 |  | 0.92  (0.82-1.00) | 80.0%  (54.8-93.0%) | 92.0%  (75.0-97.8%) |
| 1.1\*TV annulus z-score +  0.7\*MPA - 5.2 | 0.53 | 26.2 | 32.2 | 0.14 | 0.40 |  | 0.92  (0.82-1.00) | 86.7%  (62.1-96.3%) | 92.0%  (75.0-97.8%) |
| 0.7\*TV annulus mm +  0.7\*MPA - 12.9 | 0.50 | 27.6 | 33.6 | 0.15 | 0.35 |  | 0.90  (0.79-1.00) | 73.3%  (48.0-89.1%) | 84.0%  (65.3-93.6%) |
| 7.2\*TV/MV annulus ratio +  0.6\*MPA - 11.1 | 0.48 | 28.4 | 34.4 | 0.16 | 0.32 |  | 0.90  (0.79-1.00) | 73.3%  (48.0-89.1%) | 84.0%  (65.3-93.6%) |
| a The proportion of biventricular repairs correctly predicted; bThe proportion of univentricular repairs correctly predicted.  R2: Efron’s pseudo-R2; AIC: Akaike Information Criterion; BBS: Brier skill score; AUC: area under the receiver operating curve;  RV: right ventricle; LV: left ventricle; TV: tricuspid valve; MV: mitral valve; MPA: main pulmonary artery; PV: pulmonary valve. | | | | | | | | | |

Table S3: Clinical patient details including interventions, complications, mortality and outcomes at last follow-up. Prediction score and ventricular outcome have been included. \* indicates incorrect prediction using the developed score.

|  |  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Patient Number (Sex)** | **Antenatal diagnosis** | **Clinical Findings** | **Initial procedure (age)** | **Subsequent procedures (age)** | **Discriminant score (%)** | | **Ventricular outcome** | **Mortality (Age, early/late)** | **Complications and morbidity** | **Outcome at last follow-up** |
| 1 (M) | No | PA/IVS | Pulmonary valve Balloon valvuloplasty (1 day) |  | 4.42 (98.8%) | Biventricular | |  |  | Stable at transition to adult service (15yrs) |
| 2 (F) | No | PA/IVS | Pulmonary valve Balloon valvuloplasty (7 days) | Balloon valvuloplasties (6mths, 4yrs) | 1.49 (81.7%) | Biventricular | |  | T1DM (Dx age 5yrs) | Stable at transition to adult service (16yrs) |
| 3 (F) | Yes | PA/IVS, coronary artery abnormality | Right mBT Shunt (1day) | BCPC, surgical pulmonary valvotomy (9mths), Fontan (3yrs 9mths) | -1.17 (23.5%) | Univentricular | |  |  | Stable at transition to adult service (16yrs) |
| 4 (M) | Yes | PA/IVS, coronary artery sinus fistula, Prematurity | Nil |  | -4.81 (0.8%) | Univentricular | | Yes (1day, pre-intervention) | Respiratory failure in prematurity |  |
| 5 (F) | No | PA/IVS with severe RV hypoplasia, RVDCC | Right mBT Shunt (3 days) | mBT Shunt revision (4yrs) | -3.75 (2.3%) | Univentricular | | Yes (4yrs, early) | OOHCA of unclear cause |  |
| 6 (F) | Yes | PA/IVS | Right mBT Shunt (3 days) | BCPC, Surgical pulmonary valvectomy (8mths), Fontan completion (3yrs 5mths) | -3.77 (2.2%) | Univentricular | |  |  | Stable at last follow-up (8yrs) |
| 7 (F) | No | PA/IVS, 22q11 microdeletion | Nil |  | -5.24 (0.5%) | Univentricular | | Yes (24 days, pre-intervention) | Necrotising enterocolitis |  |
| 8 (M) | Yes | PA/IVS, coronary artery fistula | Right mBT Shunt (2 days) | Diagnostic angiography (8mths) | -3.56 (2.7%) | Univentricular | | Yes (12mths, late) | OOHCA of unclear cause |  |
| 9 (F) | No | PA/IVS | Pulmonary valve perforation and balloon valvuloplasty (4 days) | Pulmonary balloon valvuloplasty (4mths), Surgical pulmonary valvectomy (4yrs) | 5.20 (99.5%) | Biventricular | |  | CVA and GMFCS II Cerebral Palsy | Stable at last follow up (11yrs) |
| 10 (F) | No | PA/IVS, Prothrombin gene mutation, coronary artery fistula | Unsuccessful Pulmonary valve perforation (1 day), Right mBT Shunt (12 days) | BCPC, Pulmonary valvectomy (14mths), Balloon pulmonary valvuloplasty (3yrs) | -0.49 (38.1%) | One-and-a-half ventricle | |  | CVA with mild Right upper limb deficit | Stable at last follow-up (13yrs) |
| 11 (M) | Yes | PAIVS, RVDCC | Right mBT Shunt (2 days) | Diagnostic angiography (9mths) | -4.00 (1.8%) | Univentricular | | Yes (8mths, late) | Protein losing enteropathy, central line infection and sepsis |  |
| 12 (M) | Yes | PA/IVS, RVDCC | Right mBT Shunt (3 days) | BCPC (9mths), Fontan (3yrs 10mths) | -5.69 (0.34%) | Univentricular | |  | CVA with persistent deficit | Stable at last follow-up (11yrs) |
| 13 (F) | No | PA/IVS | Attempted pulmonary valve perforation (2days), Right mBT Shunt with pulmonary valvotomy (6 days) | Balloon pulmonary valvuloplasty (7mths), BCPC, pulmonary valvectomy (13mths) | -0.11 (47.2%) | One-and-a-half ventricle | |  |  | Stable at last follow-up (12yrs) |
| 14 (F) | No | PA/IVS | Pulmonary valve perforation and balloon valvuloplasty (3 days) |  | 1.22 (77.1%) | Biventricular | |  |  | Mild pulmonary stenosis at last follow-up (10yrs) |
| 15 (M) | Yes | PA/IVS | Right mBT Shunt (1 day) | Pulmonary valve radiofrequency perforation and balloon valvuloplasty (4mths), mBT Shunt division and pulmonary valvotomy (12mths) Balloon valvuloplasties (14mths, 2 years) | 2.59 (93.0%) | Biventricular | |  |  | Stable at last follow-up (10yrs) |
| 16 (F) | Yes | PA/IVS, coronary artery fistula, Noonan Syndrome | Right mBT Shunt (8 days) | Central mBT Shunt (7mths), BCPC, mBT shunt takedown and pulmonary valvectomy (17mths), Fontan (4yrs 8mths) | -3.82 (2.1%) | Univentricular | |  | Post-operative junctional ectopic tachycardia | Stable at last follow-up (11yrs) |
| 17 (M) | Yes | PA/IVS | Radiofrequency pulmonary valve perforation (5 days) |  | 2.24 (90.4%) | Biventricular | |  |  | Moderate pulmonary regurgitation at last follow-up (9yrs) |
| 18 (M) | Yes | PA/IVS, Left anterior descending artery stenosis | Right mBT shunt (2 days) | Diagnostic angiography (6mths) | -6.71 (0.1%) | Univentricular | | Yes (6mths, early) | Necrotising enterocolitis, sepsis post initial surgery. Significant arrhythmia post angiography at 6mths |  |
| 19 (F) | Yes | PA/IVS, RVDCC | Left mBT Shunt (2 days) | mBT shunt revision (29 days) | -5.22 (0.5%) | Univentricular | | Yes (1mth, early) | Bradycardia and arrest post operatively |  |
| 20 (M) | Yes | PA/IVS, Extreme prematurity | Nil |  | -4.96 (0.69) | Univentricular | | Yes (13 days, pre-intervention) | Necrotising enterocolitis |  |
| 21 (M) | Yes | PA/IVS, Coronary artery fistula | Right mBT Shunt and PDA ligation (4 days) | Pulmonary balloon valvuloplasty (24 days), Pulmonary surgical valve repair (14mths) | 0.26 (56.5%) | Biventricular | |  |  | Stable at last follow-up (8yrs) |
| 22 (M) | Yes | PA/IVS | Radiofrequency pulmonary valve perforation (3 days) | Pulmonary balloon valvuloplasty (6yrs 2mths) | 2.21 (90.2%) | Biventricular | |  |  | Stable at last follow-up (7yrs) |
| 23 (F) | Yes | PA/IVS, Coronary artery fistula | Percutaneous pulmonary valve perforation (2 days) |  | -3.91 (2.0%) \* | Biventricular | |  |  | Stable at last follow-up (5yrs) |
| 24 (M) | Yes | PA/IVS, Extreme prematurity, RVDCC | Nil |  | -5.39 (0.5%) | Univentricular | | Yes (14 days, pre-intervention) | Significant respiratory distress and arrest |  |
| 25 (M) | Yes | PA/IVS | Pulmonary valve radiofrequency perforation (2 days) |  | 1.62 (83.4%) | Biventricular | |  |  | Stable at last follow-up (5yrs) |
| 26 (F) | Yes | PA/IVS, Prematurity | Right mBT shunt (8 days) | RVPA conduit formation (6mths) | 0.12 (53.1%) | Biventricular | |  | Sepsis and renal failure post-operatively | Stable at last follow-up (3yrs) |
| 27 (M) | No | PA/IVS | Radiofrequency pulmonary valve perforation (2 days) | Pulmonary balloon valvuloplasty (3mths) | 3.48 (97.0%) | Biventricular | |  |  | Stable at last follow-up (3yrs) |
| 28 (M) | No | PA/IVS | Percutaneous pulmonary valve perforation (1 day) | Failed pulmonary balloon valvuloplasty and surgical valvectomy (2mths), RVPA Conduit (2yrs 6mths) | 1.67 (84.2%) | Biventricular | |  | Sepsis post-surgical valvectomy | Stable at last follow-up (3yrs) |
| 29 (F) | Yes | PA/IVS, MAPCAs | Nil |  | -3.69 (2.4%) | Univentricular | | Yes (9 days, pre-intervention) |  |  |
| 30 (M) | Yes | PA/IVS, RVDCC | Radiofrequency pulmonary valve perforation (1 day) | Right mBT Shunt (33 days), BCPC (4mths), | -3.11 (4.2%) | Univentricular | |  | OOHCA in rural centre, CVA, Renal failure necessitating dialysis | Awaiting Fontan completion at last follow-up (3yrs) |
| 31 (M) | Yes | PA/IVS, Coronary artery fistula | Right mBT Shunt (8 days) | mBT Shunt revision (3mths), RVPA Conduit (6mths), Central mBT shunt placement (6.5mths), Pulmonary artery balloon dilatation (13mths) | -7.99 (0.03%) | Univentricular | |  | Post-catheter venous thrombosis | Awaiting further surgical management at last follow-up (22mths) |
| 32 (M) | No | PA/IVS | Percutaneous Pulmonary Valve perforation (4 days) | Right mBT Shunt (7 days), BCPC and surgical valvectomy (10mths) | -3.11 (4.3%) | Univentricular | |  | Post-operative pericardial effusion | Awaiting Fontan at last follow-up (18mths) |
| 33 (F) | Yes | PA/IVS, coronary artery fistula | Radiofrequency pulmonary valve perforation (3 days) | Right mBT Shunt (7days), BCPC and surgical valvectomy (9mths) | -1.70 (15.4%) | Univentricular | |  |  | Awaiting Fontan completion at last follow-up (3yrs) |
| 34 (M) | Yes | PA/IVS | Right mBT Shunt (6 days) | Radiofrequency pulmonary valve perforation (13 days), BCPC 8mths) | -2.26 (9.4%) | Univentricular | |  |  | Awaiting Fontan completion at last follow-up (3.5yrs) |
| 35 (M) | Yes | PA/IVS | Failed percutaneous Pulmonary valve perforation (1day), Surgical valvectomy (4 days) | Balloon pulmonary valvuloplasty (3mths), Surgical pulmonary valve repair (10mths) | 8.50 (99.9%) | Biventricular | |  | Parvovirus myocarditis | Stable at transition to adult services (16yrs) |
| 36 (M) | Yes | PA/IVS | Unsuccessful radiofrequency pulmonary valve perforation (1 day) | Right mBT shunt (21 days), surgical pulmonary valvectomy (4mths) | -0.65 (34.2%) | One-and-a-half ventricle | |  | Myocardial perforation at initial catheterisation with tamponade | Awaiting BCPC at last follow up (18mths) |
| 37 (F) | No | PA/IVS, Protein C Deficiency, Right Aortic arch | Failed radiofrequency perforation (2 days), Surgical pulmonary valvectomy (8 days) | Right mBT Shunt (21 days), Central shunt formation (5mth) | 1.23 (77.4%)\* | Univentricular | | Yes (5mth, early) | Thrombosed mBT shunt, sepsis, adrenal infarction |  |
| 38 (M) | Yes | PA/IVS, Coronary artery fistula | Right mBT Shunt (1 day) | RVPA Conduit (14mths), BCPC (2yrs 8mths), Fontan (4yrs 6mths) | -5.70 (0.33%) | Univentricular | |  | Sternal wound infection | Stable at transition to adult services (16yrs) |
| 39 (M) | Yes | PA/IVS, Coronary artery fistula, Liver haemangiomas | Radiofrequency pulmonary valve perforation (2 days) | Right mBT Shunt (21 days), BCPC, (12mths) | -1.26 (22.0%) | One-and-a-half ventricle | |  |  | Stable at last follow-up (18mths) |
| 40 (F) | Yes | PA/IVS | Right mBT Shunt (1 day) | RVPA Conduit and mBT shunt takedown (9mths) | 1.42 (80.5%) | Biventricular | |  |  | Stable at latest follow-up (2yrs) |

BCPC: bidirectional cavopulmonary connection; CP: cerebral palsy; CVA: cerebral vascular accident; GMFCS: Global motor and functional classification scale; MAPCAs: major aortopulmonary collateral arteries; mBT Shunt: modified Blalock-Taussig shunt; OOHCA: out of hospital cardiac arrest; PA/IVS: pulmonary atresia with intact ventricular septum; PDA: patent ductus arteriosus; RV: right ventricle; RVDCC: right ventricle dependent coronary circulation; RVPA conduit: right ventricle to pulmonary artery conduit; T1DM: Type 1 diabetes mellitus.

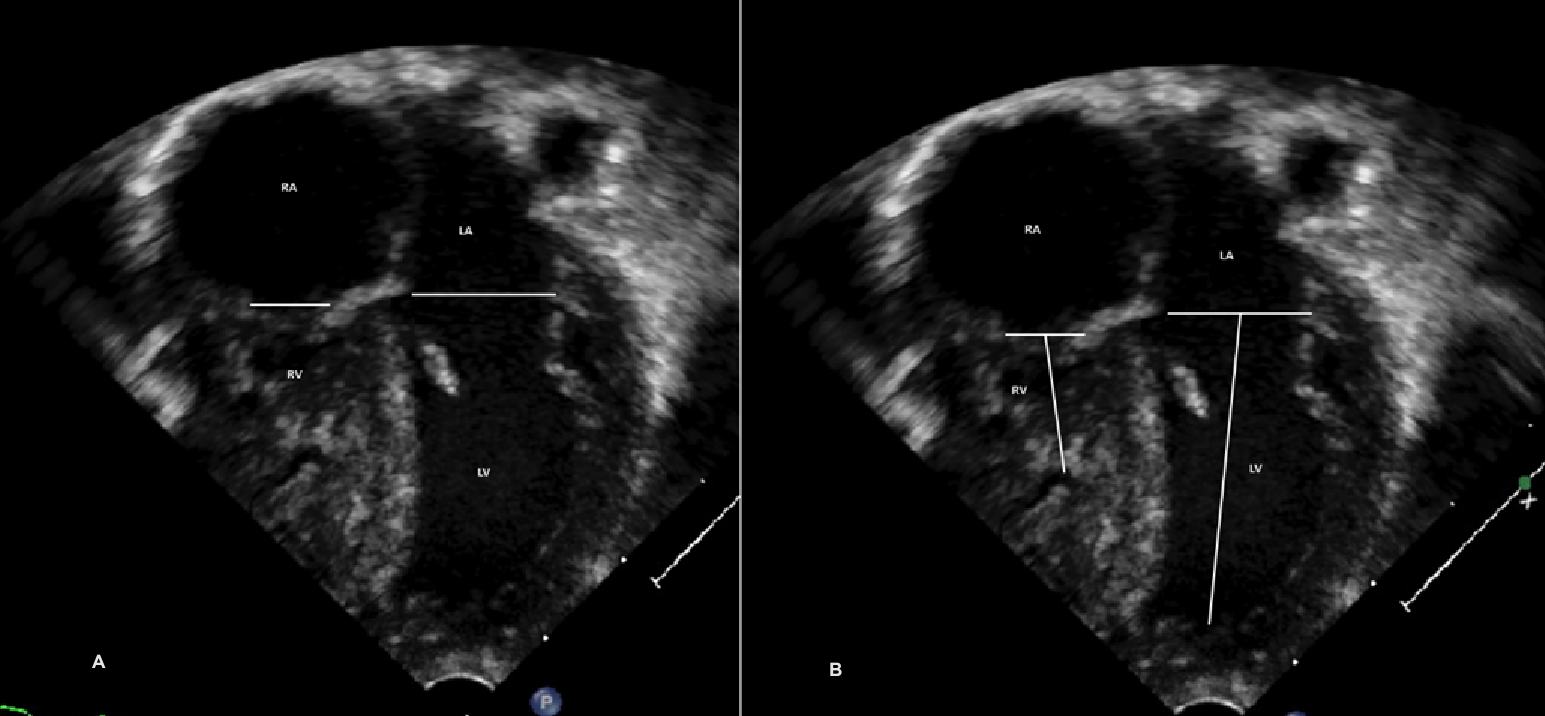


Figure S1. Transthoracic echocardiography demonstrating measurements used in the discriminatory score in an apical 4 chamber view. The echocardiogram was taken at time of diagnosis postnatally and the patient achieved biventricular repair. Image A shows tricuspid valve annulus and mitral valve annulus measurements Image B demonstrates right ventricular length and left ventricular length measurements. RA = Right Atrium, RV = Right Ventricle, LA = Left Atrium, LV = Left Ventricle.

A close-up of a ultrasound

Description automatically generated

Figure S2. Transthoracic echocardiography apical 4 chamber view of a patient at birth (A) and at latest follow-up at 8 years of age (B). At initial echocardiography, right ventricular length was 20mm, tricuspid valve annulus was 9mm, left ventricular length 26mm and mitral valve annulus was 12mm. The discriminatory score predicted the child had a 56.5% chance of being selected for biventricular repair. The patient underwent an initial modified Blalock-Taussig shunt, followed by percutaneous pulmonary valve perforation at day 24 of life. The patient achieved biventricular repair following surgical valve repair and modified Blalock-Taussig shunt take down at 15 months of age. (Video clips (1-5) are provided in supplemental material, under Case 1). RA = Right Atrium, RV = Right Ventricle, LA = Left Atrium, LV = Left Ventricle, TV Tricuspid Valve, MV = Mital Valve.



Figure S3. Transthoracic echocardiography apical 4 chamber views of a patient at birth (A) and at latest follow-up at 10 years of age (B). At initial echocardiography, right ventricular length was 19mm, tricuspid valve annulus was 11mm, left ventricular length 32mm and mitral valve annulus was 13mm. Using the discriminatory score, the child had a predicted 47.2% chance of achieving biventricular repair. The patient progressed to a one-and-a-half ventricular palliation following an initial modified Blalock-Taussig shunt at birth and later surgical pulmonary valvotomy, bidirectional cavopulmonary connection formation and modified Blalock-Taussig shunt take down at 13 months of age. (Video clips (6-9) are provided in supplemental material, under Case 2). RA = Right Atrium, RV = Right Ventricle, LA = Left Atrium, LV = Left Ventricle, TV Tricuspid Valve, MV = Mital Valve.

A collage of images of human heart

Description automatically generated

Figure S4. Three-dimensional shell and blood pool renderings from cardiac tomography angiograms of a patient at day two of life (A & C), and again at 12 months of age, pre-bidirectional cavopulmonary connection (B & D), demonstrating relative changes to the right ventricle blood pool volume. This patient underwent radiofrequency perforation of the pulmonary valve at birth followed by a modified Blalock-Taussig shunt as initial management. Using the discriminatory score, the child had a predicted chance of biventricular outcome of 34% from birth. The patient has since achieved a one-and-a-half ventricular palliation. RA = Right Atrium, RV = Right Ventricle, LV = Left Ventricle, Ao = Aorta, SVC = Superior Vena Cava, PA = Pulmonary Artery.

Movie clips

*Case 1 achieved biventricular repair, corresponding to Figure S2 in the supplemental materials.*

Clip 1: Apical 4 chamber view at initial echocardiography demonstrating significant right ventricular hypoplasia

Clip 2: Parasternal short axis view with colour Doppler across the right ventricular outflow tract at initial echocardiography showing membranous pulmonary atresia

Clip 3: Apical 4 chamber view at 12 months of age with colour Doppler demonstrating moderate tricuspid regurgitation prior to surgical pulmonary valve repair and modified Blalock-Taussig shunt take down

Clip 4: Apical 4 chamber view at latest follow-up at 8 years, showing good biventricular function with significant development of the right ventricle

Clip 5: Parasternal short axis view at latest follow-up at 8 years showing a well-developed right ventricular outflow tract

*Case 2 achieved a one-and-a-half ventricle circulation, corresponding to Figure S3 in the supplementary materials.*

Clip 6: Apical 4 chamber view at initial echocardiography compared with colour Doppler demonstrating right ventricular hypoplasia and tricuspid regurgitation

Clip 7: Parasternal short axis view at initial echocardiography showing membranous pulmonary atresia

Clip 8: Apical 4 chamber view at latest follow-up at 10 years, demonstrating increased right ventricular dimensions and good biventricular function. The patient underwent device closure of an atrial septal defect to achieve a one-and-a-half ventricular circulation

Clip 9: Parasternal short axis view with colour Doppler at latest follow-up at 10 years, demonstrating a well-developed right ventricular outflow tract with flow acceleration across the pulmonary valve and pulmonary regurgitation, post-surgical pulmonary valvotomy

**Supplementary Materials S4:**

The Perth Pulmonary Atresia Discriminant Score (PPDS) calculator aims to guide initial management of children born with pulmonary atresia with intact ventricular septum (PAIVS). Due to the heterogenous nature of the cardiac anomaly, decision making can be challenging in borderline cases.

The PPDS discriminatory calculator uses echocardiographic measurements at the first echocardiogram, to assist clinicians in their initial decision-making process.

Click the link below to open the PPDS calculator:

<https://pch-cardiology.shinyapps.io/PAIVS_ventricular_prediction/>

*Information of note prior to using the PPDS calculator*:

* The score identifies eventual surgical management based on echocardiography at birth. The model is not designed to evaluate patients at the time of second stage decision making. Standardised quantification of echocardiographic measurements should be performed according to American Society of Echocardiography recommendations.1
* The tool does not consider other anatomical factors such coronary artery anomalies or any comorbid congenital heart lesions. These must be considered when using the score to aid decision making.
* The tool is not suitable for use in any other congenital heart lesions aside from PA/IVS.
* The output from this tool provides an estimate of the chance of proceeding to biventricular repair in eligible patients, rather than treatment recommendations.

***Disclaimer:*** The PPDS calculator is designed for use by medical professionals. Use of this Score and the information/data derived from it is not a substitute for individual case-based management as guided by patient-based decision making in a multidisciplinary clinical setting. The Score is designed by the Western Australian Child and Adolescent Health Service and is intended for personal non-commercial purposes. The Score is provided “AS IS” and The Western Australian Child and Adolescent Health Service makes no other warranties with respect to the Score, including but not limited to those of merchantability and fitness for a particular purpose.

By using the Score, you hereby waive any claims, causes of action and demands against The Western Australian Child and Adolescent Health Service (including its employees, physicians, directors and agents) in any way related to use of the Score and the information/data derived from it.

Relevant publications

1. Lopez L, Saurers DL, Barker P, Cohen MS, Colan SD, Dwyer J et al. Guidelines for Performing a Comprehensive Pediatric Transthoracic Echocardiogram: Recommendations From the American Society of Echocardiography. J Am Soc Echocardiogr. 2024 Feb;37(2):119-170.

**Supplementary Materials S5:**

**Development of the discriminatory scoring tool**

Eight baseline echocardiogram parameters reported or observed to clinically correlate with surgical outcome were considered as candidate variables in the development of a discriminatory tool for prediction of biventricular repair. Complete data was available for these eight variables; pulmonary valve dimension and Z-score had been discounted from consideration as more than 15% of values were missing.

Development of the tool utilized a series of bivariable logistic regression analyses, with biventricular repair as the outcome, where the candidate parameters were jointly modelled in pairs. Relative model performance was evaluated by examination of Efron’s pseudo-, the residual deviance and Akaike. Efron’s pseudo- is the square of the correlation between predicted probabilities and a binary outcome and measures the proportion of the variability in the outcome not explained by the model. It is defined as

where here the are indicators for the observed outcome of biventricular repair, the are the predicted probabilities, and the observed proportion of individuals achieving biventricular repair.

Predictive performance was assessed by leave-one-out cross validation as a measure of internal validity. Predicted probabilities were obtained for each individual from the model trained on the dataset from which they were omitted. The Brier score, was then calculated as the mean over all leave-one-out predictions of the squared difference between predictions and observed value, and the Brier skill score as .

The ability of the scores obtained from the fitted models to discriminate surgical outcome was evaluated by examination of the area under the receiver operating curve, sensitivity and specificity. In this context, sensitivity is the proportion of correctly identified biventricular repairs and specificity is the proportion of correctly identified univentricular repairs.

A discriminatory score was derived from the coefficients of each fitted candidate model. As logistic regression models probability of outcome on the log-odds scale, a score greater than zero is taken as a predictor of biventricular repair when applied to any individual, and the corresponding predicted probability calculated as: