

Supplementary Material to:

Anatomical Severity of Ebstein's Anomaly: a Quantitative Analysis based on Cardiovascular Magnetic Resonance Imaging

CMR images acquisition protocol

Each CMR scan included: (i) retrospectively gated, breath-held, balanced steady-state free precession cine (cine-SSFP) sequences (vertical RV long-axis, full heart short-axis and horizontal long-axis stack; slice thickness, 8.0 mm; no gap; flip angle, 60°–80°; repetition time, 3.8 ms; echo time, 1.7 ms; typical readout field of view, 350 mm; phase resolution, 75%; typical acquired voxel size 1.4×1.4 mm; lines per segment, 15); (ii) 2D PC-MR sequences, the following image parameters were considered: TE = 2.47 – 2.92 ms, TR = 4.64 – 6.39 ms, slice thickness = 6 mm, pixel spacing = 1.77 – 2.08 mm, reconstructed temporal resolution = 28.5 – 38.9 ms. VENC = 150-180 cm/s to avoid aliasing, flip-angle = 20°; (iii) 2D gradient echo inversion recovery LGE images (three long-axis views, full LV short-axis stack) 8–10 min after contrast.

Reproducibility analysis

Intra- and inter-observer reproducibility of key CMR measurements was evaluated using Bland-Altman analysis and the intra-class correlation coefficient (ICC) for continuous variables, and the Cohen's Kappa coefficient for categorical variables.

Inter-observer reproducibility analysis of Carpentier classes, SLD, ILD, EVRA, and right-sided heart volumes in systole and diastole was performed in 20 patients randomly chosen by two blinded operators with five years of experience (G.P., level 3 EACVI CMR CHD certification, L.T. level 3 EACVI CMR certification). G.P. re-analyzed the same dataset weeks apart, blinded from the first measurements, for intra-observer variability.

Patient Population

Most frequent associated anomalies included atrial septal defect in 40 patients (67%), features of non-compaction cardiomyopathy in 23 patients (38%), mitral valve prolapse in 5 (8%), ventricular septal defect in 2 (3%). Hypertrophic cardiomyopathy, pulmonary valve dysplasia and bicuspid aortic valve were each observed in one subject. No patient had more than trivial pulmonary regurgitation. Two patients had undergone surgical intervention (a modified Blalock-Taussig shunt and a closure of atrial septal defect) and four other patients had a percutaneous closure of an atrial septal defect. Seventeen patients had a history of documented arrhythmias, of whom 11 were diagnosed with Wolff-Parkinson-White syndrome, while 1 patient had experienced sustained VT. At CMR examination, 21 patients (44%) exhibited symptoms of clinical decompensation, with 11 patients in NYHA class II (52%) and 10 in NYHA class III (48%). Among symptomatic patients, 5 (24%) were excluded from TV surgery: a patients had just a mild TR and underwent to isolated BPCA, while four other patients were referred to the Advanced Heart Failure Team for assessment as potential candidates for further therapeutic strategies. Among the asymptomatic patients, 14 out of 39 (36%) exhibited severe TR, with progression in fRV dilation or systolic dysfunction and a decline in predicted peak oxygen consumption at consecutive monitoring exams. Following multidisciplinary heart team discussion, these patients were subsequently recommended for surgery (**Figure 1S**).

Surgical technique

Since 2015, our center has adopted the cone repair as the standard surgical approach for patients with Ebstein's anomaly. The is based on complete delamination and mobilization of all available leaflet tissue, which is then reattached circumferentially to the true right atrioventricular junction to create a 360-degree leaflet cone. A key additional part of the procedure is the remodeling of the atrialized portion of the right ventricle. This part does not effectively contribute to systolic function, so it can be reduced through plication or resection. The extent of remodeling depends strictly on the

anatomy and course of the right coronary artery, as special care must be taken to avoid iatrogenic injury or distortion of coronary flow. In our centre, RV resection over plication is preferred, as it better preserves ventricular geometry, avoids annular distortion, and reduces the risk of coronary injury, thrombus formation, and plication dehiscence. When required, complementary annular plication is performed to reduce the tricuspid annulus to a size appropriate for the reconstructed neo-tricuspid valve. The repair may then be further stabilized with the implantation of an annuloplasty ring.

Surgical Results

Overall 30 out of 60 patients underwent TV surgery; of them, 27 out of 30 (90%) had CR, while 3 underwent TV replacement. Associated procedures included triangular resection of the aRV in 16 patients, RA plasty in 4, ring annuloplasty in 16, SL augmentation in 4 patients, and Cox-Maze procedure in two. Ventricular plication was performed in 3 patients (11%), two in the early phase of our experience and one with a previous atrial septal defect closure, in whom dense adhesions precluded safe coronary assessment for resection. In all remaining patients (24 pts – 89%), triangular resection of the atrialized right ventricle was performed.

Among patients with associated anomalies, one patient underwent pulmonary valve replacement, two patients underwent ventricular septal defect closure, and closure of an interatrial communication was performed in all but three patients. Mean cardiopulmonary bypass (CPB), aortic clamping, and total operation time were 150.9 ± 34.2 , 127.6 ± 27.0 , and 296 ± 56.4 minutes, respectively. All patients received inotropic therapy with milrinone during the ICU stay; of them 28 patients were administered adrenaline, while two patients received noradrenaline as a third inotropic drug. The mean orotracheal intubation time was 25.8 ± 22.8 hours. Of the 29 patients who were discharged to the regular ward, the mean ICU stay was 5.5 ± 3.0 days and the total hospital stay was 22.4 ± 11.4 days. Post-operative complications other than hemodynamic instability and Cone breakdown included pleural and pericardial effusion in 11 patients (3 of whom required surgical

drainage), and non-sustained arrhythmias (2 of supraventricular origin, one ventricular). Four out of nine patients who experienced hemodynamic instability in the ICU stay had a BCPA (44%). No significant difference was observed in significant RVD at discharge between the BCPA group (n = 8, 90%) and the patients without BCPA (n = 13, 70%, p = 0.3). **Figure 2S** displays changes in tricuspid regurgitation at admission and discharge. Mean anterograde pressure gradient across the neo-valve was registered as 3.3 ± 1.6 mmHg.

Supplementary figures

Figure S1. Study population and flow-Chart of the clinical decision making on the Study Population.

Abbreviations: BCPA, bidirectional cavo-pulmonary anastomosis, CMR, cardiovascular magnetic resonance, EA, Ebstein's Anomaly, fRVD functional right ventricular dilation or dysfunction, HF heart failure; TR tricuspid regurgitation, TV tricuspid valve.

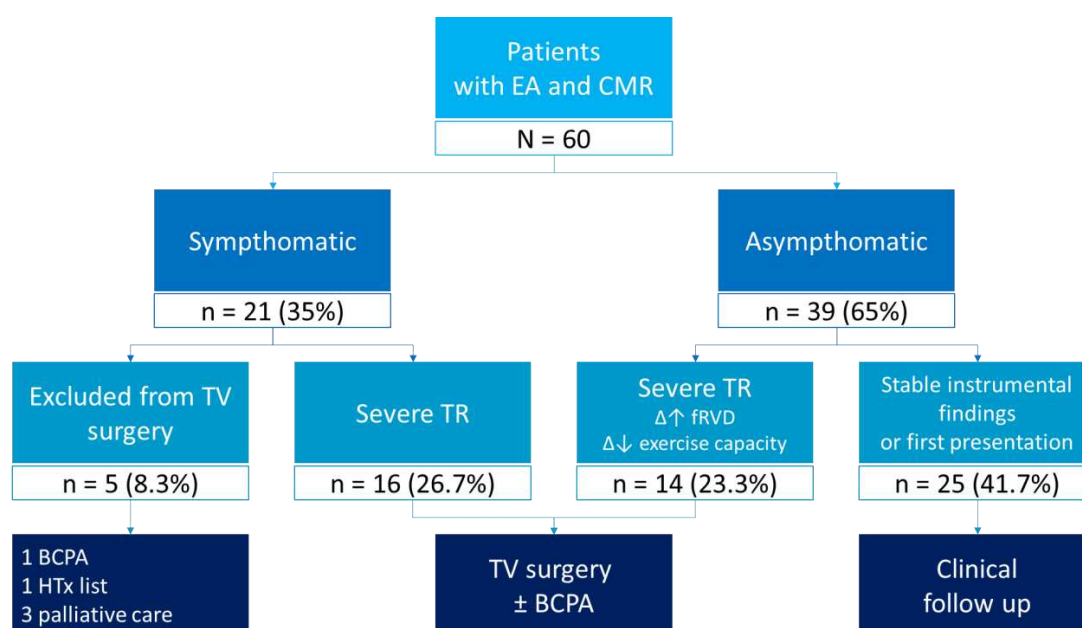
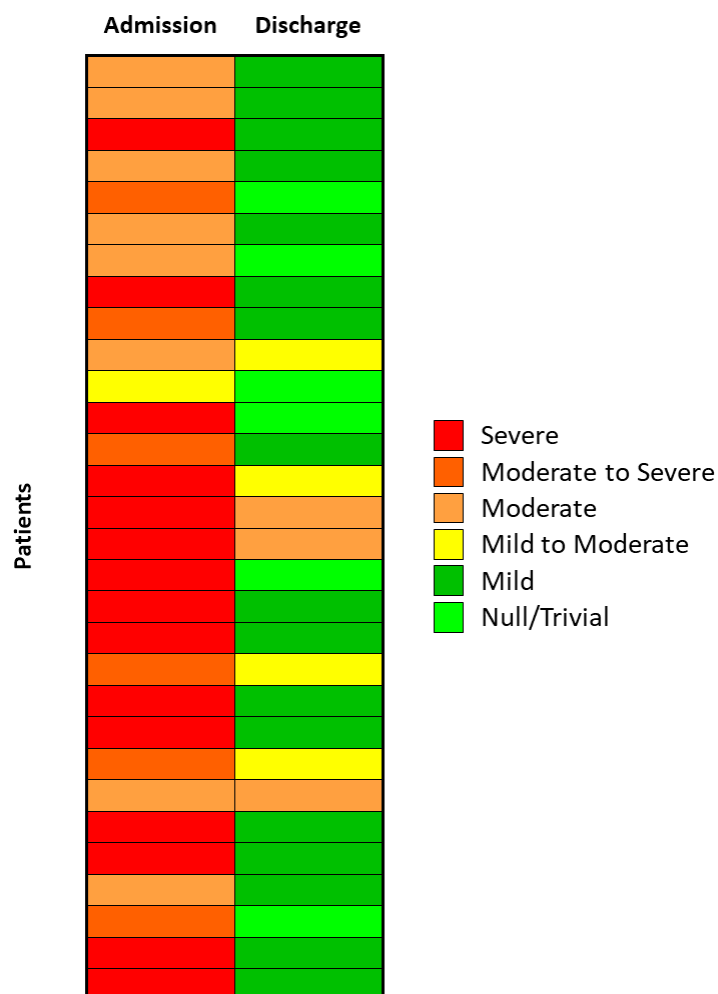


Figure S2. Tricuspid regurgitation in the surgical group assessed by transthoracic echocardiography at hospital admission and discharge.



Supplementary Tables

Table S1. Intraobserver and interobserver variability analysis.

Variable	Intraobserver (n = 20)		Interobserver (n = 20)	
	Bland-Altman	<i>r</i>	Bland-Altman	<i>r</i>
fRV EDV, mL	4.0 (-15.7, 23.7)	0.99	5.1 (-42.3, 52.4)	0.96
fRV ESV, mL	2.9 (-17.1, 22.9)	0.99	0.9 (-56.4, 58.2)	0.91
aRV EDV, mL	-0.3 (-8.0, 7.4)	0.99	-8.1 (-56.0, 39.8)	0.96
RA EDV, mL	-20.5 (-96.8, 55.9)	0.91	-5.4 (-128.2, 117.5)	0.73
RA ESV, mL	22.3 (-92.2, 136.8)	0.93	8.7 (-88.9, 106.3)	0.95
EVRA, °	1.5 (-8.9, 11.9)	0.99	1.1 (-20.7, 22.9)	0.94
SLD, mm	1.0 (-3.5, 5.4)	0.99	-1.4 (-9.4, 6.7)	0.98
ILD, mm	1.4 (-2.3, 5.0)	0.99	-0.7 (-6.7, 5.4)	0.99

Bland-Altman biases (limits of agreement) and Pearson correlation coefficient (*r*) for the intraobserver and interobserver variability analysis. Abbreviations: aRV, atrialized right ventricle, fRV, functional right ventricle, EDV, end-diastolic volume, ESV, end-systolic volume, EVRA, Ebstein's Valve Rotation Angle, ILD, inferior leaflet displacement, SLD, septal leaflet displacement, RA, right atrium.

Table S2. Multinomial logistic regression analysis of the association between each index of EA severity and the Carpentier classification.

Variable	P value	Reference Carpentier group: A					
		Carpentier group B		Carpentier group C		Carpentier group D	
		P value	OR (95% CI)	P value	OR (95% CI)	P value	OR (95% CI)
Age, years	0.76	0.67	1.01 (0.96, 1.06)	0.32	1.02 (0.98, 1.07)	0.71	1.01 (0.95, 1.07)
Male, n (%)	0.91	0.82	1.19 (0.28, 5.49)	0.72	0.77 (0.19, 3.19)	0.91	1.11 (0.16, 7.52)
Arrhythmic history, n (%)	0.001	< 0.001	E ⁹ (E ⁸ , E ¹⁰)	< 0.001	E ⁹ (E ⁸ , E ¹⁰)	< 0.001	E ¹⁰ (E ¹⁰ , E ¹⁰)
BSA, Kg/m ²	0.95	0.63	1.56 (0.26, 9.26)	0.58	1.61 (0.30, 8.49)	0.70	1.57 (0.17, 14.7)
HR, bpm	0.52	0.26	0.97 (0.92, 1.02)	0.72	0.99 (0.95, 1.04)	0.29	0.97 (0.90, 1.03)
aRV EDVi, ml/m ²	< 0.001	0.15	1.03 (0.99, 1.08)	0.005	1.06 (1.02, 1.11)	< 0.001	1.09 (1.04, 1.14)
ARV EDVi, ml/m ²	0.01	0.81	0.99 (0.98, 1.01)	0.08	1.01 (0.99, 1.03)	0.03	1.02 (1.00, 1.04)
fRV EDVi, ml/m ²	0.22	0.28	0.99 (0.97, 1.01)	0.50	1.01 (0.99, 1.02)	0.93	1.00 (0.98, 1.02)
fRV ESVi, ml/m ²	0.04	0.88	1.00 (0.97, 1.04)	0.08	1.03 (0.99, 1.06)	0.09	1.03 (0.99, 1.07)
fRV MI, g/m ²	0.18	0.05	0.94 (0.89, 1.00)	0.16	0.97 (0.93, 1.01)	0.55	0.98 (0.93, 1.04)
fRV FE, %	< 0.001	0.03	0.87 (0.76, 0.99)	0.003	0.82 (0.72, 0.94)	0.001	0.77 (0.66, 0.89)
LV EDVi, ml/m ²	0.55	0.29	1.03 (0.97, 1.10)	0.94	1.00 (0.95, 1.06)	0.46	1.03 (0.96, 1.10)
LV ESVi, ml/m ²	0.09	0.10	1.11 (0.98, 1.26)	0.21	1.08 (0.96, 1.22)	0.02	1.21 (1.03, 1.41)
LV MI, g/m ²	0.48	0.63	1.01 (0.96, 1.08)	0.45	0.98 (0.92, 1.04)	0.71	1.01 (0.94, 1.09)
LV FE, %	0.03	0.12	0.90 (0.79, 1.03)	0.04	0.88 (0.77, 0.99)	0.008	0.82 (0.71, 0.95)
LV NCF, n (%)	0.29	0.34	2.46 (0.40, 15.15)	0.13	3.80 (0.68, 21.28)	0.10	5.99 (0.70, 50.0)
TV RF, %	0.41	0.39	0.98 (0.94, 1.02)	0.64	1.01 (0.97, 1.05)	0.61	0.99 (0.94, 1.04)
ATV AP, mm	0.02	0.45	1.03 (0.96, 1.11)	0.01	1.10 (1.02, 1.19)	0.38	1.04 (0.95, 1.14)
ATV SI, mm	0.45	0.63	0.98 (0.90, 1.07)	0.74	1.01 (0.94, 1.09)	0.32	1.05 (0.96, 1.15)
RA ESVi, ml/m ²	0.13	0.88	1.00 (0.98, 1.02)	0.16	1.01 (0.99, 1.03)	0.19	1.02 (0.99, 1.03)
CO, L/min	0.63	0.97	0.99 (0.60, 1.65)	0.56	0.86 (0.53, 1.42)	0.29	0.67 (0.32, 1.41)
CI, L/min/m ²	0.45	0.41	0.63 (0.21, 1.88)	0.20	0.50 (0.18, 1.43)	0.15	0.37 (0.09, 1.45)
SLD, mm	< 0.001	0.01	1.12 (1.02, 1.22)	0.003	1.14 (1.05, 1.25)	< 0.001	1.21 (1.09, 1.34)
SLDi, mm/m ²	< 0.001	0.02	1.16 (1.03, 1.30)	0.003	1.19 (1.06, 1.34)	< 0.001	1.29 (1.13, 1.49)
ILd, n (%)	< 0.001	< 0.001	E ¹⁰ (E ⁹ , E ¹¹)	< 0.001	E ¹¹ (E ¹⁰ , E ¹²)	< 0.001	E ¹¹ (E ¹¹ , E ¹¹)

ILD , mm	< 0.001	< 0.001	2.68 (2.48, 2.89)	< 0.001	2.88 (2.71, 3.07)	< 0.001	3.06 (3.06, 3.06)
ILD_i , mm/m ²	< 0.001	< 0.001	4.45 (3.97, 4.98)	< 0.001	5.08 (4.71, 5.47)	< 0.001	5.43 (5.43, 5.43)
EVRA , °	< 0.001	< 0.001	1.80 (1.46, 2.23)	< 0.001	2.17 (1.82, 2.59)	< 0.001	2.77 (2.77, 2.77)
aRV/ARV_{2D} , -	< 0.001	0.006	E ² (4.08, E ⁴)	0.003	E ² (6.13, E ⁴)	0.001	E ² (14.8, E ⁵)
aRV/ARV_{3D} , -	< 0.001	0.04	E ³ (1.61, E ⁸)	0.005	E ⁶ (35.2, E ⁹)	< 0.001	E ¹⁰ (E ⁶ , E ¹⁶)
fRV/ARV_{2D} , -	0.002	0.64	1.26 (0.47, 3.38)	0.01	0.01 (E ⁻⁵ , 0.35)	0.007	E ⁻⁵ (E ⁻⁸ , 0.06)
fRV/ARV_{3D} , -	< 0.001	0.04	E ⁻⁴ (E ⁻⁸ , 0.62)	0.005	E ⁻⁶ (E ⁻⁹ , 0.03)	< 0.001	E ⁻¹¹ (E ⁻¹⁷ , E ⁻⁶)
fRV/LV , -	0.08	0.12	0.43 (0.15, 1.23)	0.53	1.26 (0.62, 2.57)	0.66	0.78 (0.25, 2.41)
Cel_{3D} , -	0.001	0.45	5.95 (0.06, E ²)	0.02	E ² (2.45, E ⁵)	0.004	E ³ (13.3, E ⁶)
Cel_{2D} , -	0.12	0.59	0.40 (0.02, 11.0)	0.29	4.27 (0.29, 64.1)	0.18	8.27 (0.39, E ²)
Total R/L , -	0.02	0.23	0.60 (0.26, 1.39)	0.22	1.49 (0.79, 2.81)	0.31	1.45 (0.71, 2.96)
Multinomial logistic regression using Carpentier group A as reference group. Abbreviations: aRV, atrialized right ventricle; ARV, anatomical right ventricle; ATV AP, anatomical tricuspid valve annulus measured in anterior-posterior direction; ATV SI, anatomical tricuspid valve annulus measured in superior-inferior direction; BSA, body surface area; Cel, Celemajer index; CI, cardiac index; CO, cardiac output; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; HR, heart rate; ILd, inferior direct leaflet delamination defect; ILDi, indexed inferior leaflet displacement; LV, left ventricle; MI, mass index; NCF, features of myocardial non compaction; NYHA, New York Heart Association class; RA, right atrium, SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index; TRf, tricuspid regurgitant fraction. Subscripts: 2D, based on long axis or, for Cel, on areas; 3D based on volumes.							

Table S3. Receiver operator characteristics curve analysis for CMR severity indices.

Variable	AUC	Cut-off	Sensitivity (%)	Specificity (%)	Positive predictive value (%)	Negative predictive value (%)
aRV EDVi , ml/m ²	0.77	34.2	22/32 (68.8)	19/28 (67.9)	22/31 (71.0)	19/29 (65.5)
ARV EDVi , ml/m ²	0.71	214.6	19/32 (59.4)	20/28 (71.4)	19/27 (70.3)	20/33 (60.6)
fRV ESVi , ml/m ²	0.69	80.3	16/32 (50.0)	18/28 (64.3)	16/26 (61.5)	18/34 (52.9)
fRV EF , %	0.74	41.9	22/32 (68.8)	18/28 (64.3)	22/32 (68.8)	18/28 (64.3)
LV EF , %	0.65	66.2	21/32 (65.6)	17/28 (60.7)	21/32 (65.6)	17/28 (60.7)
ATV AP , mm	0.70	49.5	24/32 (75.0)	17/28 (60.7)	24/35 (68.6)	17/25 (68.0)
SLD , mm	0.74	24.5	20/32 (62.5)	15/28 (53.6)	20/33 (60.6)	15/27 (55.6)
SLDi , mm/m ²	0.75	28.4	24/32 (75.0)	16/28 (57.1)	24/36 (66.7)	16/24 (66.7)
ILD , mm	0.89	25.5	30/32 (93.8)	23/28 (82.1)	30/35 (85.7)	23/25 (92.0)
ILDi , mm/m ²	0.89	17.8	29/32 (90.6)	22/28 (78.6)	29/35 (82.9)	22/25 (88.0)
EVRA , °	0.98	54	29/32 (90.6)	25/28 (89.3)	29/32 (90.6)	25/28 (89.3)
aRV/ARV_{2D} , -	0.74	0.99	21/32 (65.6)	21/28 (75.0)	21/28 (75.0)	21/32 (65.6)
aRV/ARV_{3D} , -	0.71	0.24	22/32 (68.8)	16/28 (57.1)	22/34 (64.7)	16/26 (61.5)
fRV/ARV_{2D} , -	0.76	0.57	26/32 (81.3)	18/28 (64.3)	26/36 (72.2)	18/24 (75.0)
fRV/ARV_{3D} , -	0.71	0.77	22/32 (68.8)	16/28 (57.1)	22/34 (64.7)	16/26 (61.5)
Cel_{3D} , -	0.73	0.46	22/32 (68.8)	17/28 (60.7)	22/33 (66.7)	17/27 (63.0)
Total R/L , -	0.70	3.10	22/32 (68.8)	17/28 (60.7)	22/33 (66.7)	17/27 (63.0)

Receiver Operating Characteristic curve analysis, including area under the curve (AUC), sensitivity, and specificity of thresholds, for indices of EA severity in mild/moderate cases (type A U type B) versus severe/extensive cases (type C U type D). Abbreviations: aRV, atrialized right ventricle; ARV, anatomical right ventricle; ATV, anatomical tricuspid valve annulus measured in anterior-posterior direction; Cel_{3D}, Celemajer index based on volumes; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; ILd, inferior direct leaflet delamination defect; ILD, inferior leaflet displacement; ILDi, indexed inferior leaflet displacement; LV, left ventricle; SLD, septal leaflet displacement; SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index. Subscripts: 2D, based on long axis; 3D based on volumes.

Table S4. Comparison of baseline characteristics and CMR features between surgical and non-surgical groups.

Variables	Surgery		P Value
	Yes (30)	No (30)	
Age, years	18 (10, 34)	23 (13, 44)	0.30
Male, n (%)	16 (53.3)	16 (53.3)	1.00
Arrhythmic history, n (%)	9 (30.0)	8 (26.7)	1.00
BSA, Kg/m ²	1.56 ± 0.47	1.66 ± 0.38	0.36
HR, bpm	74 (66, 82)	67 (57, 85)	0.17
aRV EDVi, ml/m ²	48.4 (28.2, 75.0)	43.3 (28.0, 66.0)	0.44
ARV EDVi, ml/m ²	171.2 (156.4, 254.3)	150.0 (124.3, 213.3)	0.02
fRV EDVi, ml/m ²	136.5 (101.9, 165.6)	109.1 (93.4, 187.7)	0.02
fRV ESVi, ml/m ²	63.1 (50.6, 104.9)	54.5 (41.1, 77.0)	0.03
fRV MI, g/m ²	27.8 (20.9, 39.4)	27.8 (23.3, 39.2)	0.67
fRV EF, %	45.9 ± 11.6	48.9 ± 10.0	0.28
LV EDVi, ml/m ²	62.1 ± 14.0	63.7 ± 13.2	0.65
LV ESVi, ml/m ²	24.8 ± 6.1	23.1 ± 7.6	0.36
LV MI, g/m ²	50.0 (37.9, 55.6)	53.7 (40.0, 68.8)	0.03
LV EF, %	61.5 (52.9, 66.3)	64.7 (59.6, 67.6)	0.03
LV NCF, n (%)	11 (36.7)	12 (40.0)	1.00
TV RF, %	47.8 ± 17.8	31.9 ± 16.2	0.001
ATV AP, mm	49.9 ± 10.4	48.2 ± 12.4	0.57
ATV SI, mm	47 (39, 55)	45 (41, 53)	0.99
RA ESVi, ml/m ²	84.4 (76.9, 118.9)	74.3 (54.8, 106.4)	0.11
CO, L/min	3.9 (2.7, 4.6)	4.2 (4.0, 5.6)	0.18
CI, L/min/m ²	2.7 ± 0.8	2.8 ± 0.8	0.76
SLD, mm	40.4 ± 17.2	37.1 ± 17.3	0.47
SLDi, mm/m ²	27.9 ± 13.3	22.4 ± 8.6	0.06
ILD, n (%)	20 (66.7)	16 (53.3)	0.43
ILD, mm	35 (18, 42)	29 (0, 43)	0.71
ILDi, mm/m ²	21.7 (14.1, 32.7)	19.8 (0.0, 27.8)	0.42
EVRA, °	59 (33, 72)	52 (15, 76)	0.87
aRV/ARV _{2D} , -	1.05 (0.68, 1.47)	0.81 (0.55, 1.22)	0.19
aRV/ARV _{3D} , -	0.27 ± 0.13	0.27 ± 0.12	0.99
fRV/ARV _{2D} , -	0.49 (0.40, 0.53)	0.58 (0.43, 0.71)	0.07
fRV/ARV _{3D} , -	0.73 ± 0.13	0.73 ± 0.12	0.99
fRV/LV, -	2.10 (1.50, 2.87)	1.72 (1.40, 2.08)	0.03
Cel _{3D} , -	0.48 (0.33, 0.66)	0.50 (0.43, 0.61)	1.00
Cel _{2D} , -	0.59 (0.52, 0.96)	0.58 (0.47, 0.81)	0.54
Total R/L, -	3.20 (2.34, 3.65)	2.59 (1.76, 3.19)	0.03

Mann-Whitney test or Student t test according to data normality for continuous variables. Chi-square test for categorical variables. Abbreviations: aRV, atrialized right ventricle; ARV, anatomical right ventricle; ATV, anatomical tricuspid valve annulus; AP, anterior-posterior; BSA, body surface area; Cel, Celemajer index; CI, cardiac index; CO, cardiac output; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; HR, heart rate; ILd, inferior leaflet direct delamination defect; ILD inferior leaflet displacement; ILDi, indexed inferior leaflet displacement; LV, left ventricle; MI, mass index; NCF, features of myocardial non compaction; RA, right atrium; SI, superior-inferior; SLD septal leaflet displacement; SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index; TRf, tricuspid regurgitant fraction. Subscripts: 2D, based on long axis or, for Cel, on areas; 3D based on volumes.

Table S5. Comparison of baseline characteristics and CMR parameters in relation to the need of a BCPA.

Variables	BCPA			Univariate	
	No (n = 21)	Yes (n = 9)	P Value	OR (95% CI)	P Value
Age, years	23 (13, 34)	13 (8, 41)	0.37	0.98 (0.93, 1.03)	0.39
Male, n (%)	13 (61.9)	3 (33.3)	0.24	3.25 (0.63, 16.8)	0.16
Arrhythmic history, n (%)	5 (23.8)	3 (33.3)	0.67	1.60 (0.29, 8.86)	0.59
BSA, Kg/m ²	1.63 ± 0.46	1.38 ± 0.47	0.19	0.29 (0.05, 1.77)	0.18
HR, bpm	72 (62, 82)	75 (70, 86)	0.21	1.04 (0.98, 1.11)	0.15
aRV EDVi, ml/m ²	43.5 (24.0, 72.6)	53.7 (39.4, 70.5)	0.53	1.01 (0.99, 1.03)	0.42
ARV EDVi, ml/m ²	182.1 (156.4, 236.4)	174.5 (163.3, 283.7)	0.86	1.00 (0.99, 1.02)	0.49
fRV EDVi, ml/m ²	138.7 (109.2, 176.3)	130.1 (104.1, 166.5)	0.63	1.00 (0.99, 1.02)	0.79
fRV ESVi, ml/m ²	73.4 (49.8, 98.9)	59.7 (51.7, 106.7)	0.69	1.01 (0.99, 1.03)	0.29
fRV MI, g/m ²	30.1 (22.2, 45.4)	24.4 (19.4, 38.8)	0.26	0.96 (0.89, 1.03)	0.25
fRV EF, %	48.4 ± 9.7	40.0 ± 14.2	0.14	0.94 (0.87, 1.01)	0.08
LV EDVi, ml/m ²	64.3 ± 12.6	57.0 ± 16.4	0.25	0.96 (0.90, 1.02)	0.19
LV ESVi, ml/m ²	25.2 (19.1, 29.3)	26.8 (20.0, 27.7)	0.72	0.96 (0.84, 1.10)	0.56
LV MI, g/m ²	50.7 (41.3, 55.8)	49.7 (37.4, 54.5)	0.45	0.97 (0.89, 1.06)	0.46
LV EF, %	60.6 ± 9.5	57.4 ± 9.5	0.38	0.95 (0.86, 1.05)	0.31
LV NCF, n (%)	7 (33.3)	5 (55.6)	0.42	2.50 (0.51, 12.4)	0.26
TV RF, %	47.0 ± 17.3	49.4 ± 19.9	0.76	1.01 (0.96, 1.06)	0.73
ATV AP, mm	51 ± 10	47 ± 10	0.28	0.96 (0.88, 1.04)	0.26
ATV SI, mm	50 ± 11	41 ± 9	0.04	0.91 (0.81, 1.01)	0.07
RA ESVi, ml/m ²	106.5 (68.6, 144.1)	83.2 (77.6, 126.7)	0.86	0.99 (0.98, 1.01)	0.74
CO, L/min	4.6 ± 1.9	3.4 ± 1.1	0.04	0.56 (0.28, 1.14)	0.11
CI, L/min/m ²	2.8 ± 0.7	2.6 ± 1.0	0.70	0.79 (0.28, 2.17)	0.64
SLD, mm	38.6 ± 18.3	44.6 ± 14.3	0.39	1.02 (0.98, 1.07)	0.38
SLDi, mm/m ²	24.9 ± 12.6	35.0 ± 12.7	0.07	1.06 (0.99, 1.14)	0.07
ILd, n (%)	11 (52.4)	9 (100.0)	0.01	E ⁻¹⁰ (0.00, n.a.)	0.99
ILD, mm	28 (0, 39)	38 (30, 66)	0.045	1.07 (1.01, 1.14)	0.02
ILDi, mm/m ²	18.2 (0.0, 23.0)	39.3 (21.0, 44.6)	0.002	1.15 (1.03, 1.28)	0.01
EVRA, °	35 (0, 65)	74 (61, 81)	0.001	1.08 (1.01, 1.15)	0.02
aRV/ARV _{2D} , -	0.86 (0.52, 1.37)	1.09 (0.97, 2.72)	0.11	2.56 (0.90, 7.29)	0.08
aRV/ARV _{3D} , -	0.27 (0.17, 0.34)	0.25 (0.19, 0.41)	0.72	10.5 (0.02, E ³)	0.47
fRV/ARV _{2D} , -	0.51 ± 0.12	0.44 ± 0.17	0.28	0.02 (0.00, 9.62)	0.21
fRV/ARV _{3D} , -	0.74 ± 0.12	0.70 ± 0.13	0.49	0.10 (0.00, 54.5)	0.47
fRV/LV, -	2.11 (1.67, 2.75)	2.74 (1.54, 3.14)	0.42	1.61 (0.75, 3.46)	0.22
Cel _{3D} , -	0.55 (0.32, 0.63)	0.47 (0.39, 0.81)	0.59	2.88 (0.11, 72.7)	0.52
Cel _{2D} , -	0.56 (0.52, 0.78)	0.59 (0.49, 1.20)	0.53	8.78 (0.53, E ²)	0.13
Total R/L, -	3.20 (2.40, 3.59)	3.57 (2.34, 4.78)	0.33	1.42 (0.83, 2.43)	0.21

Mann-Whitney test or Student t test according to data normality for continuous variables. Chi-square test for categorical variables. Abbreviations: aRV, atrial right ventricle; ARV, anatomical right ventricle; ATV, anatomical tricuspid valve annulus; AP, anterior-posterior; BSA, body surface area; Cel, Celemajer index; CI, cardiac index; CO, cardiac output; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; HR, heart rate; ILd, inferior leaflet direct delamination defect; ILD inferior leaflet displacement; ILDi, indexed inferior leaflet displacement; LV, left ventricle; MI, mass index; NCF, features of myocardial non compaction; RA, right atrium; SI, superior-inferior; SLD septal leaflet displacement; SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index; TRf, tricuspid regurgitant fraction. Subscripts: 2D, based on long axis or, for Cel, on areas; 3D based on volumes.

Table S6. Comparison of baseline characteristics and CMR parameters in relation to the occurrence of hemodynamic instability in the post-operative stay.

Variables	Hemodynamic conditions			Univariate	
	Stable (n = 21)	Unstable (n = 9)	P Value	OR (95% CI)	P Value
Age, years	19 (13, 34)	23 (8, 45)	0.86	1.00 (0.96, 1.05)	0.88
Male, n (%)	11 (52.4)	5 (55.6)	1.00	0.88 (0.18, 4.23)	0.87
Arrhythmic history, n (%)	3 (14.3)	5 (55.6)	0.03	7.50 (1.25, 45.2)	0.03
BSA, Kg/m ²	1.61 ± 0.42	1.43 ± 0.57	0.33	0.41 (0.07, 2.35)	0.32
HR, bpm	74 ± 12	78 ± 22	0.49	1.02 (0.97, 1.07)	0.48
aRV EDVi, ml/m ²	42.7 (24.0, 68.8)	63.1 (47.9, 85.0)	0.13	1.01 (0.99, 1.04)	0.22
ARV EDVi, ml/m ²	194.5 ± 52.2	203.4 ± 84.1	0.78	1.00 (0.99, 1.02)	0.72
fRV EDVi, ml/m ²	145.2 (120.1, 174.2)	112.3 (91.0, 167.3)	0.16	0.99 (0.98, 1.01)	0.55
fRV ESVi, ml/m ²	74.3 (57.7, 105.6)	52.3 (45.0, 93.7)	0.23	1.00 (0.98, 1.02)	0.98
fRV MI, g/m ²	27.6 (20.6, 43.4)	31.8 (25.6, 40.6)	0.33	1.02 (0.97, 1.08)	0.47
fRV FE, %	46.4 ± 11.7	44.5 ± 12.0	0.69	0.99 (0.92, 1.06)	0.68
LV EDVi, ml/m ²	64.0 ± 12.6	57.8 ± 16.8	0.27	0.97 (0.91, 1.03)	0.27
LV ESVi, ml/m ²	26.8 (19.4, 29.1)	25.9 (19.8, 28.3)	0.76	0.98 (0.86, 1.12)	0.76
LV MI, g/m ²	50.1 ± 8.4	44.2 ± 9.7	0.10	0.93 (0.84, 1.02)	0.11
LV FE, %	60.6 ± 7.1	57.4 ± 10.0	0.33	0.95 (0.86, 1.05)	0.32
LV NCF, n (%)	8 (38.1)	4 (44.4)	1.00	1.30 (0.27, 6.33)	0.75
TV RF, %	47.8 ± 13.5	47.7 ± 26.4	0.99	1.00 (0.96, 1.05)	0.98
ATV AP, mm	50 ± 8	49 ± 15	0.72	0.98 (0.91, 1.06)	0.64
ATV SI, mm	48 ± 9	47 ± 15	0.84	0.99 (0.92, 1.07)	0.83
RA ESVi, ml/m ²	104.0 (78.1, 118.0)	84.6 (60.6, 233.2)	0.98	1.01 (0.99, 1.02)	0.27
CO, L/min	4.3 (3.4, 5.3)	3.0 (2.4, 4.1)	0.08	0.53 (0.25, 1.10)	0.09
CI, L/min/m ²	2.8 (2.4, 3.5)	2.0 (1.8, 3.5)	0.19	0.59 (0.20, 1.71)	0.33
SLD, mm	38.4 ± 18.7	44.9 ± 12.8	0.36	1.02 (0.98, 1.07)	0.34
SLDi, mm/m ²	24.9 ± 12.8	34.9 ± 12.3	0.06	1.06 (0.99, 1.13)	0.07
ILd, n (%)	11 (52.4)	9 (100.0)	0.01	E ⁻¹⁰ (0.00, n.a.)	0.99
ILD, mm	28 (0, 40)	37 (31, 51)	0.10	1.04 (0.99, 1.09)	0.08
ILDi, mm/m ²	19.8 (0.0, 23.5)	34.3 (17.8, 42.7)	0.03	1.09 (1.01, 1.18)	0.02
EVRA, °	40 (0, 72)	63 (45, 76)	0.21	1.03 (0.99, 1.07)	0.11
aRV/ARV _{2D} , -	0.89 (0.52, 1.24)	1.42 (0.88, 2.19)	0.07	2.13 (0.79, 5.75)	0.14
aRV/ARV _{3D} , -	0.24 ± 0.11	0.35 ± 0.13	0.03	E ³ (0.95, E ⁷)	0.052
fRV/ARV _{2D} , -	0.51 ± 0.13	0.46 ± 0.17	0.41	0.07 (0.00, 30.5)	0.07
fRV/ARV _{3D} , -	0.76 ± 0.11	0.65 ± 0.13	0.03	0.01 (0.00, 1.06)	0.052
fRV/LV, -	2.15 (1.87, 2.79)	1.96 (1.35, 3.27)	0.69	1.22 (0.62, 2.40)	0.57
Cel _{3D} , volumes	0.47 ± 0.20	0.68 ± 0.27	0.02	73.6 (0.99, E ³)	0.05
Cel _{2D} , areas	0.55 (0.50, 0.71)	0.94 (0.59, 1.01)	0.04	18.7 (0.87, E ²)	0.06
Total R/L, -	3.20 (2.42, 3.63)	3.57 (2.19, 4.40)	0.66	1.31 (0.82, 2.08)	0.26

Mann-Whitney test or Student t test according to data normality for continuous variables. Chi-square test for categorical variables. Abbreviations: aRV, atrialized right ventricle; ARV, anatomical right ventricle; ATV, anatomical tricuspid valve annulus; AP, anterior-posterior; BSA, body surface area; Cel, Celemajer index; CI, cardiac index; CO, cardiac output; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; HR, heart rate; ILd, inferior leaflet direct delamination defect; ILD inferior leaflet displacement; ILDi, indexed inferior leaflet displacement; LV, left ventricle; MI, mass index; NCF, features of myocardial non compaction; RA, right atrium; SI, superior-inferior; SLD septal leaflet displacement; SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index; TRf, tricuspid regurgitant fraction. Subscripts: 2D, based on long axis or, for Cel, on areas; 3D based on volumes.

Table S7. Comparison of baseline characteristics and CMR parameters in relation to the severity of right ventricle dysfunction at discharge after surgical repair.

Variables	Significant RVD at discharge			Univariate	
	No (n = 8)	Yes (n = 21)	P Value	OR (95% CI)	P Value
Age, years	22 ± 14	26 ± 18	0.46	1.02 (0.97, 1.07)	0.49
Male, n (%)	5 (55.6)	11 (52.4)	0.99	1.14 (0.24, 5.46)	0.87
Arrhythmic history, n (%)	1 (11.1)	7 (33.3)	0.37	0.25 (0.03, 2.42)	0.23
BSA, Kg/m ²	1.58 ± 0.51	1.55 ± 0.46	0.87	0.85 (0.16, 4.69)	0.85
HR, bpm	69 (59, 85)	75 (68, 81)	0.42	1.03 (0.97, 1.10)	0.34
aRV EDVi, ml/m ²	42.7 (31.5, 56.5)	53.7 (22.6, 76.0)	0.53	1.01 (0.99, 1.04)	0.33
ARV EDVi, ml/m ²	181.7 ± 38.3	203.8 ± 69.5	0.28	1.01 (0.99, 1.02)	0.37
fRV EDVi, ml/m ²	136.3 ± 28.6	143.4 ± 50.2	0.63	1.00 (0.99, 1.02)	0.68
fRV ESVi, ml/m ²	58.1 (45.8, 88.3)	74.5 (54.6, 109.2)	0.28	1.01 (0.99, 1.04)	0.32
fRV MI, g/m ²	25.0 (21.7, 38.1)	30.1 (21.1, 44.1)	0.59	1.02 (0.96, 1.09)	0.46
fRV FE, %	57.6 (43.2, 58.7)	43.6 (38.2, 53.5)	0.06	0.92 (0.84, 1.01)	0.08
LV EDVi, ml/m ²	63.4 ± 13.7	61.6 ± 14.4	0.74	0.99 (0.94, 1.05)	0.73
LV ESVi, ml/m ²	23.1 ± 6.1	25.5 ± 6.1	0.33	1.07 (0.94, 1.23)	0.31
LV MI, g/m ²	49.2 (37.7, 53.8)	53.6 (38.3, 55.8)	0.40	1.04 (0.95, 1.13)	0.41
LV FE, %	66.1 (59.8, 68.0)	59.3 (51.0, 63.2)	0.08	0.89 (0.78, 1.02)	0.09
LV NCF, n (%)	3 (33.3)	9 (42.9)	0.70	0.67 (0.13, 3.41)	0.63
TV RF, %	49.4 ± 9.1	47.0 ± 20.6	0.74	0.99 (0.95, 1.04)	0.73
ATV AP, mm	51 ± 9	50 ± 11	0.72	0.99 (0.91, 1.07)	0.73
ATV SI, mm	44 ± 7	49 ± 12	0.21	1.05 (0.96, 1.15)	0.31
RA ESVi, ml/m ²	106.5 (72.2, 126.5)	84.6 (73.2, 144.3)	0.89	1.00 (0.99, 1.02)	0.61
CO, L/min	3.9 (3.1, 4.9)	3.9 (2.7, 5.0)	0.79	0.89 (0.57, 1.38)	0.61
CI, L/min/m ²	2.8 ± 0.7	2.7 ± 0.9	0.71	0.84 (0.31, 2.26)	0.73
SLD, mm	33 ± 14	43 ± 18	0.10	1.04 (0.99, 1.10)	0.14
SLDi, mm/m ²	21.1 ± 5.2	30.9 ± 14.6	0.07	1.07 (0.99, 1.16)	0.08
ILD, n (%)	5 (55.6)	15 (71.4)	0.43	0.50 (0.10, 2.53)	0.40
ILD, mm	24 (0, 37)	37 (22, 43)	0.09	1.04 (1.00, 1.09)	0.07
ILDi, mm/m ²	14.1 (0.0, 20.5)	23.0 (16.5, 36.8)	0.02	1.08 (1.01, 1.17)	0.03
EVRA, °	32 (0, 72)	55 (35, 72)	0.23	1.02 (0.99, 1.05)	0.13
aRV/ARV _{2D} , -	0.74 (0.62, 1.06)	1.09 (0.62, 1.92)	0.11	3.44 (0.70, 16.6)	0.13
aRV/ARV _{3D} , -	0.25 ± 0.07	0.28 ± 0.14	0.51	10.1 (0.01, E ³)	0.50
fRV/ARV _{2D} , -	0.51 (0.44, 0.54)	0.49 (0.38, 0.61)	0.53	0.16 (E ⁻³ , 51.0)	0.53
fRV/ARV _{3D} , -	0.75 ± 0.07	0.72 ± 0.14	0.51	0.10 (E ⁻³ , 75.1)	0.50
fRV/LV, -	2.15 (1.72, 2.71)	2.11 (1.55, 2.92)	0.79	1.27 (0.56, 2.89)	0.58
Cel _{3D} , volumes	0.46 ± 0.16	0.56 ± 0.26	0.21	8.99 (0.15, E ²)	0.29
Cel _{2D} , areas	0.55 (0.52, 0.76)	0.62 (0.51, 0.98)	0.50	5.50 (0.18, E ²)	0.33
Total R/L, -	3.22 (2.29, 3.61)	3.28 (2.37, 3.67)	0.69	1.20 (0.68, 2.14)	0.53

Mann-Whitney test or Student t test according to data normality for continuous variables. Chi-square test for categorical variables. Significant RVD dysfunction: ≥ moderate as assessed by transthoracic echocardiography. Abbreviations: aRV, atrialized right ventricle; ARV, anatomical right ventricle; ATV, anatomical tricuspid valve annulus; AP, anterior-posterior; BSA, body surface area; Cel, Celemajer index; CI, cardiac index; CO, cardiac output; EDVi, indexed end-diastolic volume; EF, ejection fraction; ESVi, indexed end-systolic volume; EVRA, Ebstein's valve rotation angle; fRV, functional right ventricle; HR, heart rate; ILd, inferior leaflet direct delamination defect; ILD inferior leaflet displacement; ILDi, indexed inferior leaflet displacement; LV, left ventricle; MI, mass index; NCF, features of myocardial non compaction; RA, right atrium; SI, superior-inferior; SLD septal leaflet displacement; SLDi, indexed septal leaflet displacement; Total R/L, total right over left volumes index; TRf, tricuspid regurgitant fraction. Subscripts: 2D, based on long axis or, for Cel, on areas; 3D based on volumes.