Appendices

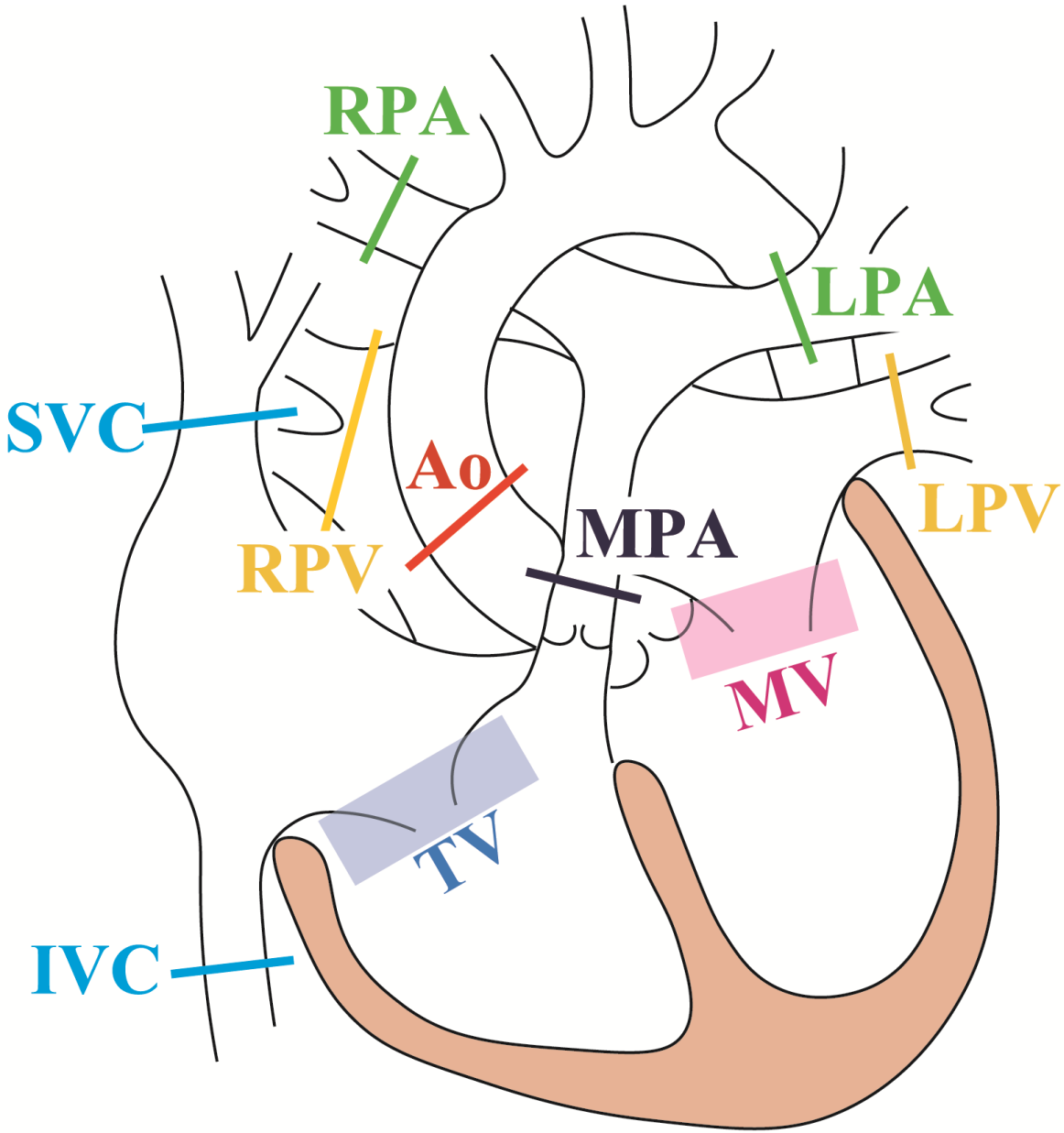
Supplemental table S1. The compatibility of quantitative analysis in CMR

	Value (L/min/m^2)	SD
Volumetric analysis		
sRVCI	**3.12**	**0.60**
pLVCI	**3.25**	**0.74**
Flow analysis		
Qs based on AV flow	**2.88**	**0.57**
Qp based on PV flow	**2.79**	**0.63**
RV in flow	**2.82**	**0.82**
LV in flow	**2.83**	**0.79**
Systemic venous return		
SVC	1.00	0.24
IVC	1.84	0.35
(Sum)	**2.84**	**0.55**
Pulmonary venous return		
RPV	1.59	0.68
LPV	1.25	0.43
(Sum)	**2.84**	**0.55**
Pulmonary artery		
RPA	1.76	0.73
LPA	1.05	0.48
(Sum)	**2.81**	**0.82**

SD, standard deviation; sRVCI, systemic right ventricle cardiac index; pLVCI, pulmonary left ventricle cardiac index; Qs, systemic blood flow; Qp, pulmonary blood flow; AV, aortic valve; PV, pulmonary valve; RV, right ventricle; LV, left ventricle; SVC, superior vena cava; IVC, inferior vena cava; RPV, right pulmonary vein; LPV, left pulmonary vein; RPA, right pulmonary artery; LPA, left pulmonary artery (The average values of each

anatomical area, N=17)

Supplemental figure S1



1. Ao, Ascending aorta
2. MPA, Main pulmonary artery
3. SVC, Superior vena cava
4. IVC, Inferior vena cava
5. RPA, Right pulmonary artery
6. LPA, Left pulmonary artery
7. MV, Mitral valve
8. TV, Tricuspid valve
9. RPV, Right pulmonary vein
10. LPV, Left pulmonary vein

Supplemental figure S1. Routine scanning of phase contrast flow by CMR.

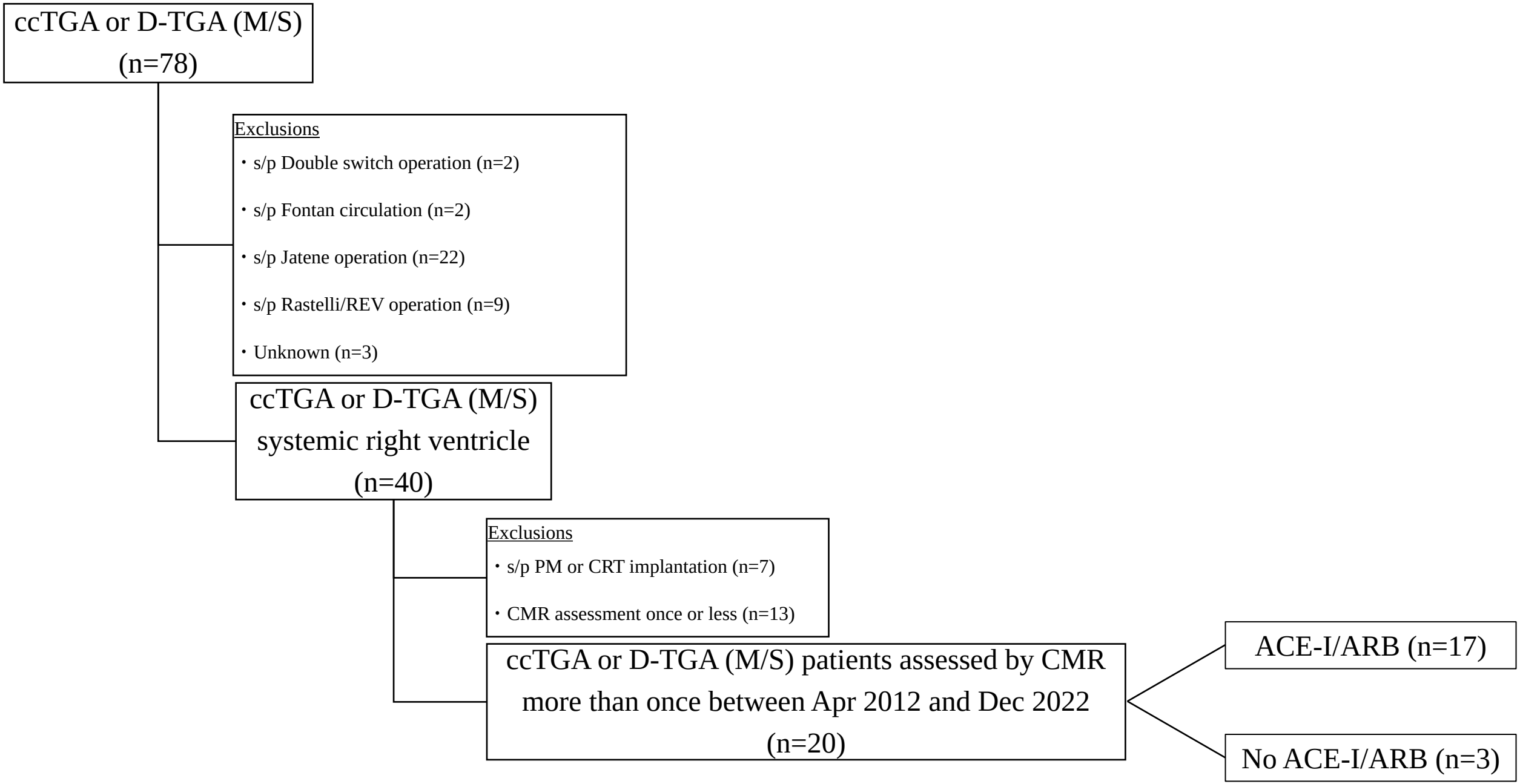
For structurally normal heart, blood flows were calculated from recordings of ten anatomical areas shown in this figure. In our study, we applied the similar methods to calculate the blood flows for patients with systemic right ventricle. In both congenitally corrected transposition of the great arteries and dextro-transposition of the great arteries after Mustard/Senning operation group, cardiac magnetic resonance (CMR) analysis was based on not functional but anatomical nomenclature.

Note that in normal hearts, a following formula is always established:

Flow 1 (ascending aorta) = Flow 2 (main pulmonary artery) = Flow 7 (mitral valve) = Flow 8 (tricuspid valve) = Flow 3+4 (sum of superior and inferior vena cava) = Flow 5+6 (sum of right and left pulmonary artery) = Flow 9+10 (sum of right and left pulmonary vein)

(Coronary flow is ignored in this formula.)

Supplemental figure S2

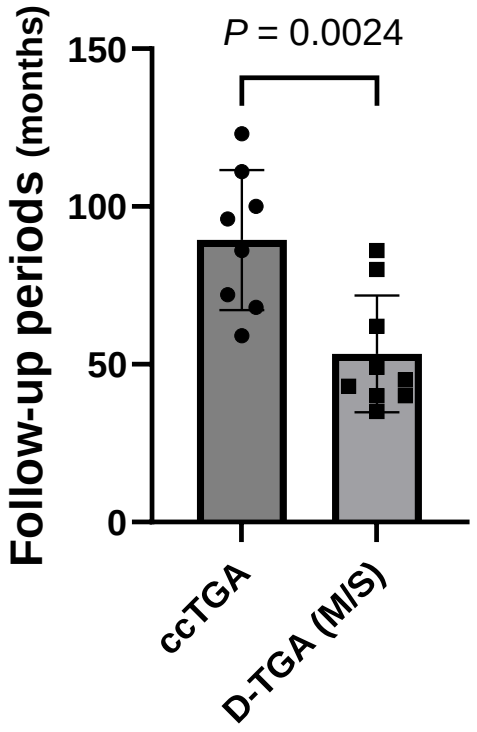


Supplemental figure S2. Flow chart of eligible patient selection

Only patients with systemic right ventricle were included in congenitally corrected transposition of the great arteries (ccTGA) and dextro-transposition of the great arteries after Mustard/Senning operation (D-TGA (M/S)) patients. After excluding patients of device implantation or assessed by cardiac magnetic resonance (CMR) once or less, twenty patients were recruited in our analysis. Among them, patients treated with ACE-I/ARB were seventeen, and the remaining did not use ACE-I/ARB.

PM, pacemaker; CRT, cardiac resynchronization therapy; REV, Rastelli/ Réparation à l'Etage Ventriculaire

Supplemental figure S3

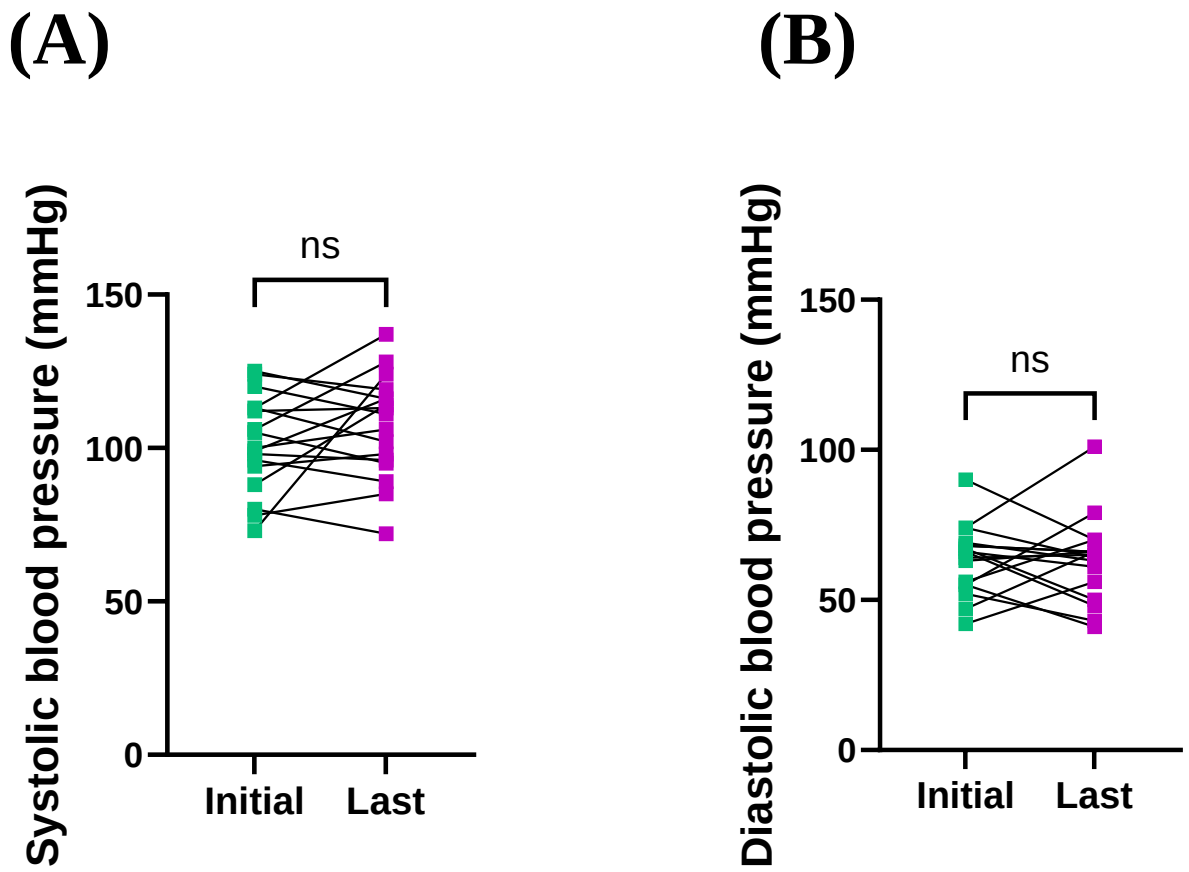


	ccTGA (n=8)	D-TGA (M/S) (n=9)	All patients (n=17)
Average CMR interval periods±SD (range), months	89.4 ± 20.8 (59-123)	53.3 ± 17.4 (35-86)	70.3 ± 26.2 (35-123)

Supplemental figure S3. Follow-up periods in the ccTGA and D-TGA (M/S) group.

The follow-up period in the ccTGA group was significantly longer than that of D-TGA (M/S) 89.4 months (range, 59-123) versus 53.3 months (range, 35-86) (N=8 for ccTGA and N=9 for D-TGA (M/S), Student's t-test, $P = 0.0024$).

Supplemental figure S4

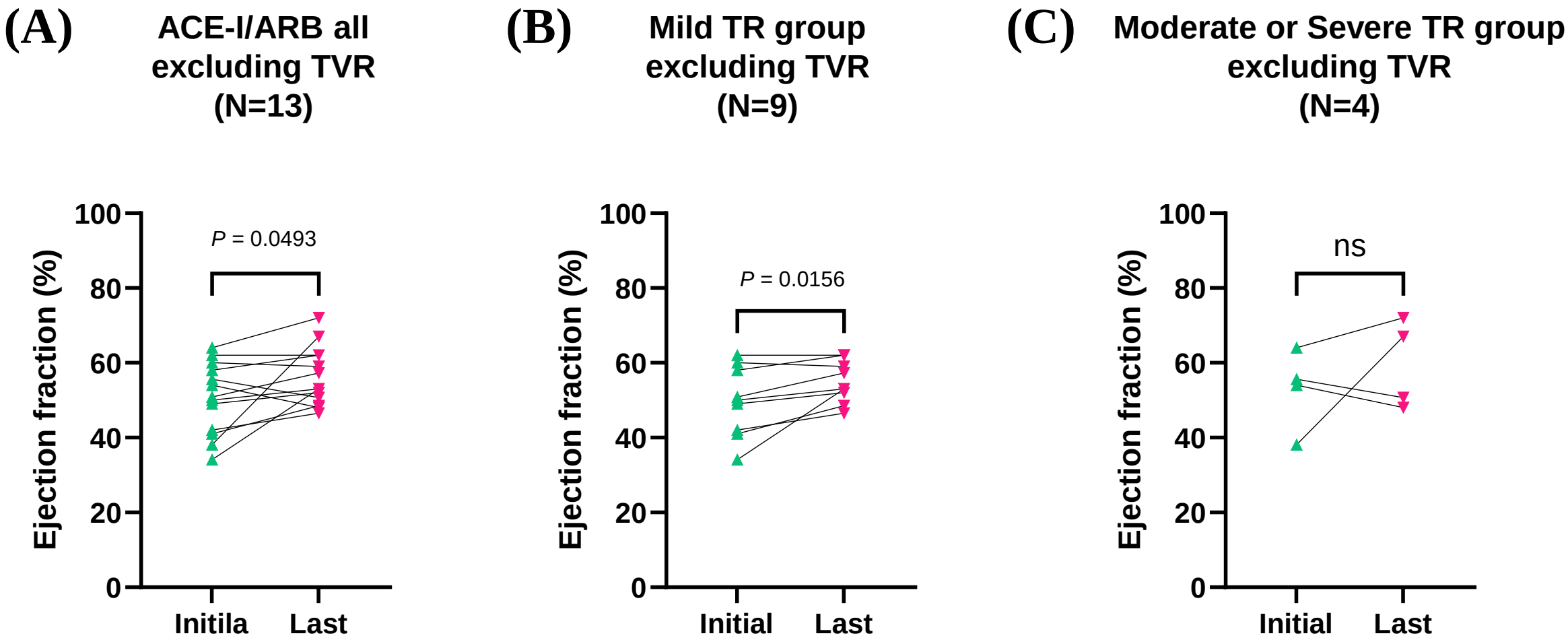


Supplemental figure S4. Changes of blood pressure between Initial and Last CMR assessments.

(A, B) There were no differences in systolic and diastolic blood pressure between Initial and Last CMR assessments.

(N=17 for all patients, Paired t-tests.)

Supplemental figure S5

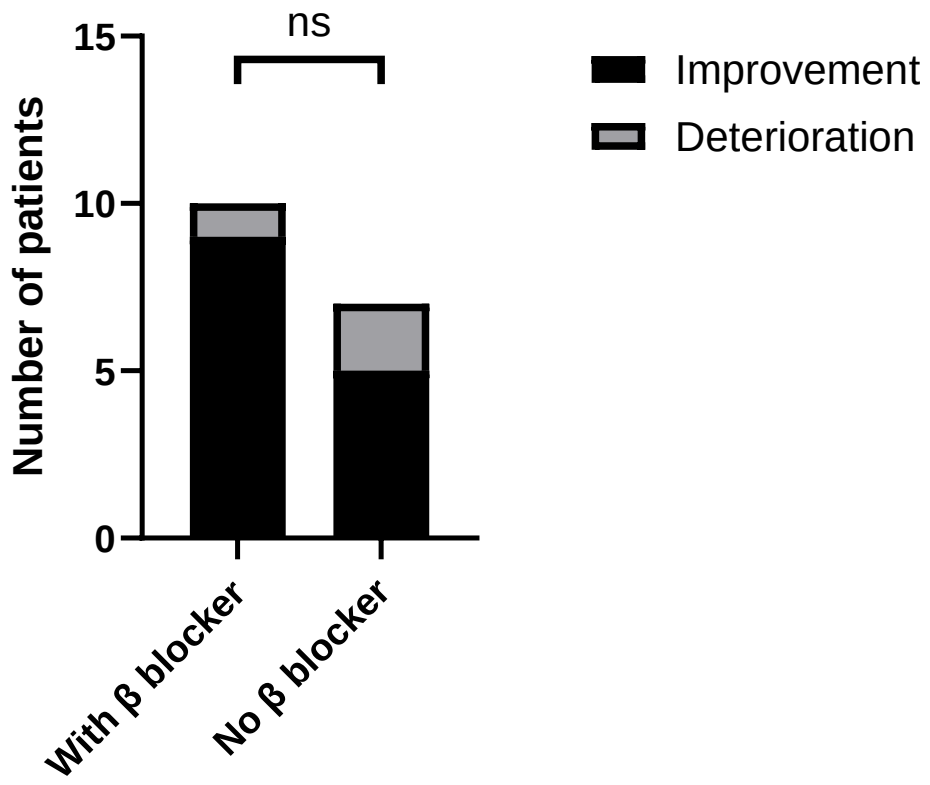


Supplemental figure S5. Changes in systemic right ventricle ejection fraction excluding patients of tricuspid valve replacement.

(A) Systemic right ventricle ejection fraction (sRVEF) improved in all patients treated with ACE-I/ARB excluding tricuspid valve replacement cases from 50.7 ± 9.5% to 56.2 ± 7.8% (N=13, Wilcoxon matched-pairs signed rank test, $P = 0.0493$). The patients were divided into two groups according to the severity of tricuspid regurgitation (TR). **(B)** The mild TR group showed significant improvement of sRVEF from 49.7±8.4% to 54.8±5.6% (N=9, Wilcoxon matched-pairs signed rank test, $P = 0.0156$), while **(C)** there was no sRVEF improvement in the moderate or severe TR group (from 52.9±10.8% to 59.4±11.9%; N=4, Wilcoxon matched-pairs signed rank test, $P = 0.6250$).

Ns, not significant

Supplemental figure S6

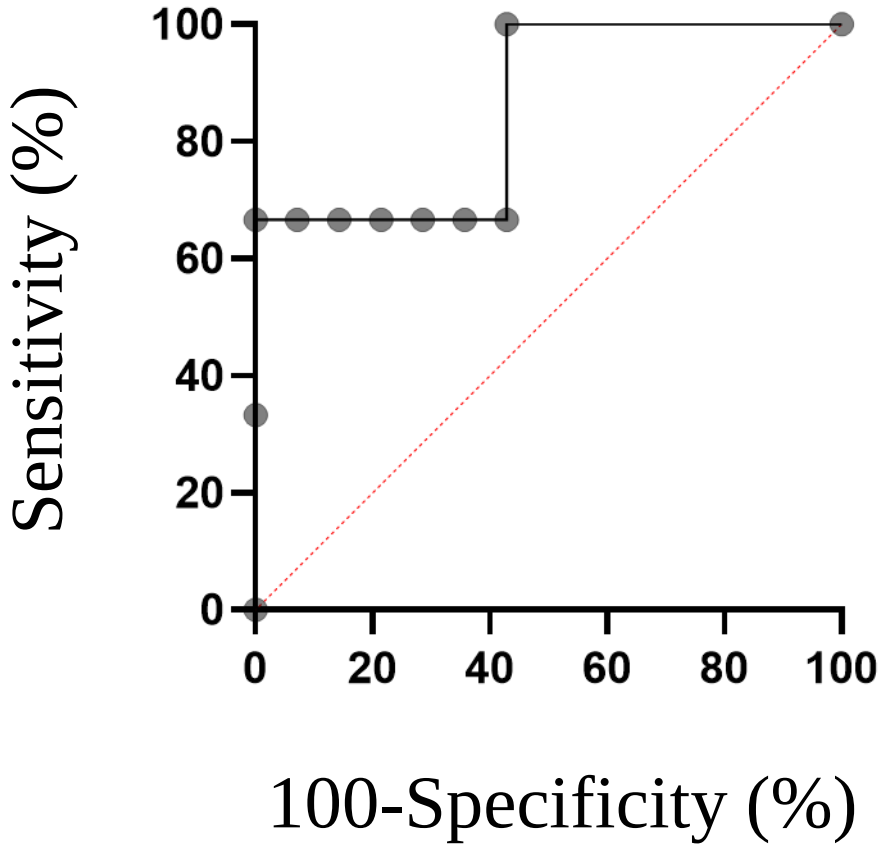


Supplemental figure S6. Comparisons of sRV function between patients with and without β blocker in our cohort.

Among ten patients who took β-blockers, nine (90%) experienced improvement in sRVEF. In the group that did not take β-blockers, five out of seven patients (71.4%) showed improvement in sRVEF. (Fisher's exact test, $P = 0.5368$)

ns, not significant

Supplemental figure S7



Tricuspid regurgitation (Cutoff value)	Area under the curve (95% CI)	*P* value	Sensitivity	Specificity
21.6%	0.8571 (0.6028-1.0000)	0.0588	66.7%	92.9%

Supplemental figure S7. Receiver operating characteristics (ROC) curve in tricuspid regurgitant fraction to predict sRVEF deterioration.

The cutoff value of 21.6% in tricuspid regurgitant fraction showed a good performance to predict systemic right ventricle ejection fraction (sRVEF) deterioration. The area under the curve (AUC) was 0.8571, and sensitivity and specificity were 66.7% and 92.9%, respectively ($P = 0.0588$).