

SUPPLEMENTAL MATERIAL

Table S1. Standard deviations of variables included in univariable Cox regression, for interpretative purposes of standardized hazard ratios.

	Standard deviation
Clinical variables	
Age, years	9
Heart rate, beats/min	13
Systolic blood pressure, mmHg	14
QRS duration, ms	21
Blood biomarkers	
	<i>(log)</i> ↓*
NT-proBNP, pmol/L	1.34
Hs-troponin-T, ng/L	1.39
GDF-15, ng/L	0.77
Hs-CRP, mg/L	1.57
Galectin-3, ng/mL	0.39
RDW, %	0.11
eGFR, mL/min/1.73m ²	0.17
Hemoglobin, mmolFe/L	0.15
Echocardiographic markers	
<i>sRV dimensions</i>	
sRV end-diastolic annulus, mm/m ²	8.6
<i>Strain parameters</i>	
LS of the sRV free wall, %	4.0
LS of the sRV septal wall, %	3.3
GCS of the sRV, %	3.8
<i>LV dimensions</i>	
LV end-diastolic diameter, mm/m ²	4.0

*Blood biomarkers were log transformed before standardization and therefore the SD's reported belong to the log transformed blood biomarker levels. CRP= C-reactive protein, eGFR= estimated glomerular filtration rate, GDF-15= growth differentiation factor-15, GCS= global circumferential strain, Hs= high sensitive, LS= longitudinal strain, LV= left ventricular, NT-proBNP= N-terminal pro b-type natriuretic peptide, RDW= red cell distribution width, sRV= systemic right ventr

Table S2. Associations between clinical variables, blood- and echocardiographic markers and the primary- (death or heart failure) and secondary endpoint (death or arrhythmia), with adjustment for New York Heart Association functional class in adults with a systemic right ventricle.

	Analysis with adjustment for NYHA functional class					
	Primary endpoint			Secondary endpoint		
	HR*	95%CI	p-value	HR*	95%CI	p-value
Clinical characteristics						
Age	1.04	0.38-1.58	0.86	0.95	0.66-1.37	0.80
Heart rate	1.47	0.89-2.44	0.14	1.48	1.02-2.13	0.038
Systolic blood pressure	0.65	0.39-1.09	0.10	0.55	0.35-0.87	0.009
QRS duration	1.23	0.80-1.91	0.35	1.30	0.92-1.83	0.13
Loss of sinus rhythm	2.16	0.85-5.56	0.11	1.61	0.75-3.47	0.23
Blood biomarkers						
NT-proBNP	1.79	1.15-2.80	0.010	1.08	0.73-1.61	0.70
Hs-troponin-T	1.70	1.03-2.78	0.036	1.40	0.96-2.03	0.08
GDF-15	2.24	1.49-3.35	<0.001	1.86	1.35-2.56	<0.001
Hs-CRP	2.02	1.18-3.44	0.010	1.38	0.92-2.06	0.12
Galectin-3	1.30	0.80-2.11	0.29	1.45	0.98-2.14	0.06
RDW	1.91	1.38-2.65	<0.001	1.71	1.26-2.31	0.001
eGFR	0.73	0.52-1.03	0.07	0.72	0.53-0.97	0.032
Hemoglobin	0.52	0.36-0.73	<0.001	0.72	0.53-1.00	0.047
Echocardiographic markers						
Tricuspid regurgitation	1.74	1.11-2.74	0.016	1.23	0.85-1.79	0.28
<i>sRV dimensions</i>						
sRV end-diastolic annulus	1.66	0.97-2.86	0.07	0.94	0.62-1.40	0.74
<i>Strain parameters</i>						
LS of the sRV free wall	1.19	0.73-1.94	0.50	1.06	0.71-1.58	0.74
LS of the sRV septal wall	0.87	0.54-1.41	0.58	0.65	0.43-0.98	0.039
GCS of the sRV [†]	-‡	-	-	0.64	0.42-0.98	0.038
<i>LV dimensions</i>						
LV end-diastolic diameter [†]	-‡	-	-	1.41	0.98-2.04	0.07

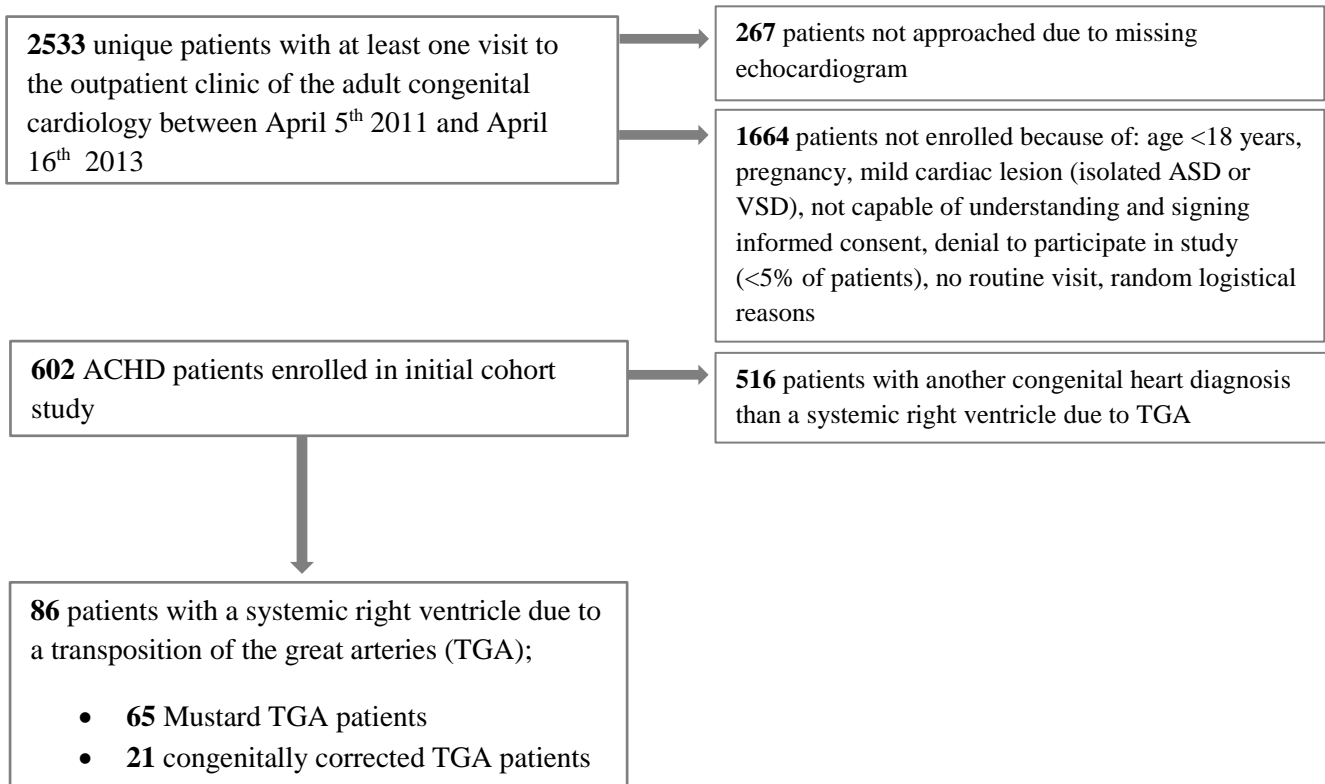
*Hazard ratios are expressed per increase in standard deviation for continuous variables. † In adult patients with transposition of the great arteries operated by the Mustard procedure only. ‡ Not enough power to perform multivariable analysis. CRP= C-reactive protein, eGFR= estimated glomerular filtration rate, GDF-15= growth differentiation factor-15, GCS= global circumferential strain, GLS= global longitudinal strain, Hs= high sensitive, LS= longitudinal strain, LV= left ventricular, NT-proBNP= N-terminal pro b-type natriuretic peptide, NYHA= New York Heart Association, RDW= red cell distribution width, sRV= systemic right ventricle

Table S3. Associations between clinical variables, blood- and echocardiographic markers and the primary and secondary endpoint, restricted to adult patients with transposition of the great arteries operated by the Mustard procedure.

	Mustard TGA patients (n=64)					
	Primary endpoint (n=15)			Secondary endpoint (n=22)		
	HR*	95%CI	p-value	HR*	95%CI	p-value
Clinical variables						
Age	2.54	1.32-4.90	0.005	2.70	1.48-4.93	0.001
Heart rate	1.48	0.87-2.52	0.15	1.53	1.03-2.28	0.036
Systolic blood pressure	0.53	0.30-0.93	0.027	0.46	0.27-0.79	0.005
NYHA >1	5.25	1.89-14.53	0.001	3.87	1.62-9.27	0.002
QRS duration	1.53	0.97-2.42	0.07	1.50	1.02-2.20	0.042
Loss of sinus rhythm	3.53	1.28-9.80	0.015	2.75	1.15-6.62	0.023
Blood biomarkers						
NT-proBNP	2.85	1.75-4.64	<0.001	1.87	1.24-2.83	0.003
Hs-troponin-T	2.58	1.55-4.31	<0.001	2.10	1.40-3.32	<0.001
GDF-15	2.47	1.61-3.78	<0.001	2.81	1.92-4.12	<0.001
Hs-CRP	2.31	1.31-4.07	0.004	1.74	1.13-2.68	0.012
Galectin-3	2.27	1.31-3.93	0.003	2.50	1.51-4.14	<0.001
RDW	2.07	1.48-2.89	<0.001	2.53	1.64-4.04	<0.001
eGFR	0.60	0.44-0.83	0.002	0.52	0.37-0.74	<0.001
Hemoglobin	0.49	0.34-0.72	<0.001	0.65	0.44-0.96	0.030
Echocardiographic markers						
Tricuspid regurgitation	1.60	0.96-2.68	0.074	1.23	0.78-1.95	0.38
<i>sRV dimensions</i>						
sRV end-diastolic annulus	2.00	0.99-4.05	0.05	1.25	0.68-2.31	0.47
<i>Strain parameters</i>						
LS of the sRV free wall	0.62	0.32-1.19	0.15	0.71	0.42-1.20	0.20
LS of the sRV septal wall	0.73	0.44-1.22	0.23	0.62	0.39-0.98	0.039
GCS of the sRV	0.77	0.46-1.30	0.33	0.69	0.46-1.05	0.09
<i>LV dimensions</i>						
LV end-diastolic diameter [#]	1.95	1.34-2.85	0.001	1.70	1.21-2.38	0.002

*Hazard ratios expressed per increase in standard deviation for continuous variables. [#]Indexed for body surface area. CRP= C-reactive protein, eGFR= estimated glomerular filtration rate, GDF-15= growth differentiation factor-15, GCS= global circumferential strain, Hs= high sensitive, LS= longitudinal strain, LV= left ventricular, NT-proBNP= N-terminal pro b-type natriuretic peptide, NYHA= New York Heart Association, RDW= red cell distribution width, sRV= systemic right ventricle

Figure S1. Flowchart of the patient selection process of the prospective observational cohort study consisting of adults with a systemic right ventricle.



ASD= atrial septal defect, VSD= ventricular septal defect, ACHD= adult congenital heart disease, TGA= transposition of the great arteries