Long term follow-up after biventricular repair of pulmonary atresia with intact ventricular septum and critical pulmonary valve stenosis.

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PURPOSE: Long term outcomes of repaired pulmonary atresia with intact ventricular septum (r*PAIVS*) and critical pulmonary valve stenosis (rCPVS) are not established. Residual lesions such as atrial septal defect, pulmonary and tricuspid regurgitation (PR and TR) may lead to right ventricle (RV) enlargement.

We sought to determine long term follow-up of patients with rPAIVS or rCPVS.

METHODS: We retrospectively studied patients with biventricular rPAIVS or rCPVS attending our cardiac magnetic resonance (CMR) imaging department. Numbers are expressed as median [interquartile range]. Pearson correlation analysis and X^2 test were used to assess the relationships between different parameters.

RESULTS: 11 patients were studied at a median age of 13.2 years [10.3-16.2]. Patients' characteristics are summearise in Table 1. Four patients had atrial septal defect requiring surgery. Age at repair was 10 days [2.5-22.7] and delay between repair and CMR study was 13.1 years [9.9-16.2]. RV was dilated 9 patients 133 ml/m² [110-164] of which 6 patients had decrease RV ejection fraction. RV volume, ejection fraction and QRS duration were normal in 2 patients. More than mild TR was present in 7 patients. All but 1 patient had PR (32% [30-39]). Late gadolinium enhancement was found in 3 patients, at infundibular level in 2. All patients had normal left ventricle volume and function. QRS duration was ≥120 ms in 5 patients with right bundle branch. RV dilatation was associated with age at CMR (r=0.62, p=0.04), decrease RV ejection fraction (r=0.78, p=0.006), a trend with TR (r=0.57, p=0.06) but not with PR (r=0.38, p=0.2). QRS duration was not associated with the type of repair, the presence late gadolinium enhancement or RV dilatation (p=0.8, p=0.4 and p=0.5 respectively).

Table 1: Patients' characteristics and CMR findings.

	Diagnosis	Age at intervention (days)	Intervention	Age at CMR (years)	TR	RVtd volume indexed (ml/m²)	RVEF (%)	LVtd volume indexed (ml/m²)	LVEF (%)	PR (%)	LGE	QRS (ms)
1	CPS	7	percututaneous dilatation	16,00	3	210		87		22	-	120
2	PAIVS	14	TAP + Blalock anastomosis	10,00	1	73	66	46	68	34	+	80
3	CPS	1	percututaneous dilatation	9,00	2	107	58		60	32		80
4	CPS	19	percututaneous dilatation	11.3	3	133	45	75	59	30		120
5	APSI	136	Blalock anastomosis	24.7	4	179	42	64	59	45		120
6	CPS	9	TAP + Blalock anastomosis	17.4	2	129	56	66	67	30	+	80
7	CPS	1	percututaneous dilatation + Blalock anastomosis	13.8	0	159	45	93	61	45	-	80
8	CPS	24	TAP	9.6	3	112	41	68	60	30	+	130
9	CPS	10	percututaneous dilatation	13.2	0	68	69	78	75	0	-	80
10	CPS	1	percututaneous dilatation	16.3	3	157	32	61	53	36	-	120
11	CPS	12	percututaneous dilatation	12.2	1	107	45	74	58	40	-	80

Legend:
CPS : Critical pulmonary stenosis ; PAIVS: pulmonary atresia with intact ventricular septum; TAP: transannular patch; CMR: cardiac magnetic resonance imaging; TR: tricuspid regurgitation; RVtd: right ventricule telediastolic; RVEF: right ventricle ejection fraction; LVtd: left ventricule telediastolic; LVEF: left ventricle ejection fraction; PR: pulmonary regurgitation; LGE: late gadolinium enhancement.

Figure 1: CMR imaging of patient n°10.

A: Cine SSFP sagital view of the right ventricle outflow track, note the valvular incompetency (arrow).

B: 4 chamber view showing RV enlargement and hypertrophy.

Legend: LV: left ventricle; RV: right ventricle.

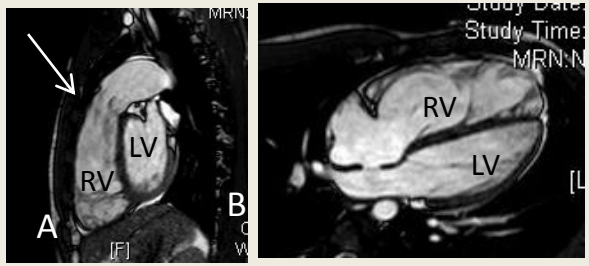
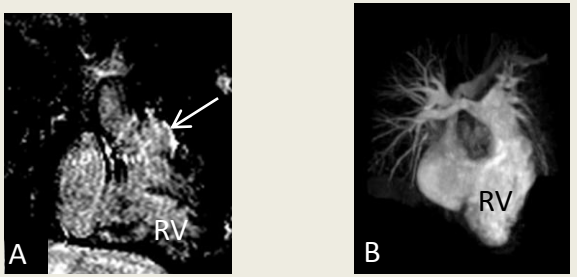


Figure 2: CMR imaging of patient n°2.

A: late infundibular guadolinium enhancement (arrow)

B: Maximum intensity projection in imaging of the rignt ventricular outflow track. Note the thickening of the apex.



CONCLUSIONS: RV dilatation, decrease ejection fraction and QRS enlargment in rPAIVS and rCPVS are common. Mechanisms seem to be multifactorial with PR and TR on an abnormal myocardium.T1 mapping studies should be performed . Determining the optimal timing for pulmonary valve replacement and tricuspid valve surgery is challenging.

Disclosures: none