Long-term outcome after surgical repair for partial anomalous pulmonary venous connection compared to isolated atrial septal defect closure.

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Introduction

Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital heart disease defined by some but not all pulmonary veins aberrantly connected to a systemic vein or to the right atrium with or without an associated atrial septal defect (ASD). The hemodynamic consequences of PAPVC are comparable to a simple secundum ASD. The only curative treatment of PAPVC is a surgical correction, whereas a secundum ASD can also be closed by a percutaneous intervention. This study aims to compare the long-term outcome between adult patients after PAPVC repair and both groups of patients with either surgical or interventionally closed simple secundum ASD (sASD, iASD, respectively), focusing on arrhythmias.

Methods

Clinical, surgical, imaging and invasive data were retrospectively reviewed from 9 centers in Austria, France and Switzerland.

Results

A total of 129 corrected PAPVC, 52 sASD and 71 iASD patients were included (for pre-repair patients' characteristics see table 1). Eighteen percent of patients with PAPVC had an intact atrial septum. sASD patients were diagnosed and operated at younger age than both other groups. PAPVC had a higher Qp:Qs preoperatively than iASD. On last follow-up, PAPVC patients presented a diminished RV longitudinal function (table 2). Exercise capacity, need for cardiac medication and prevalence of symptoms were not different between groups (p>0.1). Supraventricular tachycardia, but not ventricular arrhythmias, were significantly more prevalent in sASD than iASD (table 2). Atrioventricular conduction disorders were significantly more frequent in PAPVC patients (table 2). PAPVC patients required significantly more often an electrophysiological study or a pacemaker implantation than iASD patients (table 2).

Conclusion

Patients requiring surgical repair of PAPVC and sASD display a higher arrhythmia burden than patients corrected by an interventional procedure. This finding is suggestive of a role played by a higher pre-repair left to right shunt and/or the surgery itself. A significant number of surgical corrected patients need electrophysiological studies and pacemaker implantation.

Table 1. Patients' characteristics before correction

	PAVPC (N= 129)	**sASD (N=52)**	**iASD (N=71)**	**p**
Women, N (%)	70 (54)	27 (52)	41 (58)	0.841
ASD, N (%)	106 (82)			
Sinus venosus type	82 (64)			
Ostium secundum	17 (14) *	52 (100)	71 (100)	<0.001
Number of veins anomaly connected				
1 vein	47 (36)			
2 veins	68 (53)			
3 veins	8 (6)			
Qp : Qs	2.5†	2.4	1.7	0.002
Pulmonary arterial hypertension	21 (16)	7 (10)	8 (15)	0.448
Age at diagnosis, years	26 ± 20‡	16 ± 14	27 ± 20	0.006
Age at correction, years	28 ± 20§	18 ± 20	29 ± 20	0.010

Data are mean ± standard deviation or n (%)

*p = 0.001 PAPVC vs. sASD and iASD, †p = 0.002 PAPVC vs. iASD; ‡p = 0.022 PAPVC vs. sASD; § p < 0.05 between groups

Table 2. Latest follow-up

	PAVPC (N= 129)	sASD (N=52)	iASD (N=71)	p
Age at latest follow-up, years	39 ± 17	35 ± 20	37 ± 18	0.405
Time since correction, years	12 ± 17*	24 ± 25	6 ± 5	<0.001
Last echocardiography				
Left ventricular systolic fraction ejection, %	63 ± 6	61 ± 5	61 ± 6	0.087
Right ventricular dilatation	46 (36)	16 (31)	17 (24)	0.255
S' wave cm/s	9.5 ± 2.3†	10.7 ±2.2	11.7 ± 2.8	<0.001
Tricuspid annular plane systolic excursion, mm	17 ± 5‡	18 ± 4	22 ± 6	<0.001
Right ventricular dysfunction	19 (15)	4 (9)	4 (6)	0.102
Valvulopathy	24 (19)	14 (27)	17 (24)	0.403
Pulmonary artery pressure, mmHg	27 ± 9	25 ± 9	28 ± 8	0.518
Pulmonary hypertension	4 (3)	2 (4)	2 (3)	0.947
Last exercise test				
Heart beat rate, % predicted	90 ± 13	91 ± 12	86 ± 17	0.594
MET, % predicted	95 ± 35	101 ± 25	94 ± 31	0.741
Prevalence of arrhythmia				
Tachyarrhythmia	47 (36)	26 (50)	15 (21)§	0.004
Supraventricular tachycardia (atrial fibrillation, flutter or tachycardia)	40 (31)	23 (44)¶	15 (21)	0.024
Ventricular tachycardia/premature beats	5 (4)	3 (6)	1 (1)	0.421
Bradycardia/Atrioventricular conduction disorder	11 (9)‖	0 (0)	1 (1)	0.015
Electrophysiological study/pacemaker implantation	23 (18) #	8 (15)	3 (4)	0.033
Surgical reintervention	18 (6)	2 (3.8)	4 (6)	0.047

Data are mean ± standard deviation or n (%) if not otherwise stated

*p < 0.05 between groups, † p <0.001 PAPVC vs. iASD; ‡ p <0.001 PAPVC vs. iASD; § p < 0.03 iASD vs. PAPVC and iASD vs. sASD, ‖ p < 0.04 PAPVC vs. sASD and iASD, ¶ p = 0.010 sASD vs. iASD, # p = 0.0075 PAPVC to iASD