# Comparison of the long-term outcome of adult patients with partial anomalous pulmonary venous connection and patients with secundum atrial septal defect.

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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 is a surgical correction; however, data on the outcome of these patients is scarce. This study aims to investigate the long-term outcome of adult patients after PAPVC repair in comparison to patients with corrected simple secundum ASD, focusing especially 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 patients were identified with corrected PAPVC and 129 patients with a corrected ASD (for patients' characteristics see table1). Eighteen percent of patients with PAPVC had an intact atrial septum. PAPVC patients were diagnosed later than ASD patients and presented with

a significantly higher Qp:Qs before correction (table 1). Fifty-eight percent of the ASD were closed percutaneously and the rest surgically. On last follow-up, PAPVC patients presented more often a dilated right ventricle (RV) and a diminished RV longitudinal function than ASD patients (table 2). Exercise capacity and prevalence of symptoms were not different between groups (table 2). The number of patients on medication in the PAPVC group was significantly higher than the ASD group with no differences in the type of medication they used. Conduction arrhythmia occurred significantly more in the PAPVC-group. The later more often needed pacemaker implantation (table 2).

*Conclusion*

Patients after PAPVC repair present more often residual RV dilatation and dysfunction. Although symptoms and exercise capacity is comparable, our study reveals a significantly higher prevalence of bradyarrhythmias in operated patients for PAPVC which often necessitated a pacemaker implantation.

Table 1. Characteristics before operation

| | **PAVPC (N= 129)** | **ASD (N= 129)** | ***P*** |
|---|---|---|---|
| Women, N (%) | 70 (54) | 71 (55) | 0.790 |
| ASD, N (%) | 106 (82) | 129 (100) | 0.000 |
| Sinus venosus type | 82 (64) | 0 | 0.000 |
| Ostium secundum | 17 (14) | 123 (95) | 0.000 |
| | | | |
| **Persistent left SVC** | 21 (16) | 2 | 0.000 |
| Number of veins anomaly connected | | | |
| 1 vein | 47 (36) | | |
| 2 veins | 68 (53) | | |
| 3 veins | 8 (6) | | |
| Valvulopathy | 20 (16) | 19 (15) | 0.862 |
| Pulmonary hypertension | 21 (16) | 15 (12) | 0.281 |
| | | | |
| Tachyarrhythmia | 6 (5) | 11 (9) | 0.210 |
| Bradyarrhythmia | 2 (2) | 1 (1) | 0.561 |
| RV dilated | 91 (71) | 55 (43) | 0.000 |
| RV systolic dysfunction | 5 (4) | 2 (2) | 0.536 |
| Symptomatic | 75 (58) | 37 (29) | 0.000 |
| Dyspnea | 65 (50) | 27 (21) | 0.000 |
| Palpitation | 10 (8) | 11 (9) | 0.820 |
| Thoracic pain | 6 (5) | 6 (5) | 1.000 |
| Syncope | 3 (2) | 3 (2) | 1.000 |
| **Qp:Qs** | 2.5 ± 1.2 | 1.9 ± 0.8 | 0.049 |
| RV size ml/m2 | 152 ± 59 | 107 ± 19 | 0.267 |
| Age at diagnosis years | 27 ± 21 | 23± 18 | 0.035 |
| Age at correction years | 28 ± 20 | 25 ± 20 | 0.542 |

Data are mean ± standard deviation or n (%)

Table 2. Latest follow-up

| | PAVPC (N= 129) | ASD (N= 129) | *P* |
|---|---|---|---|
| Age at latest follow-up, years | 39 ± 17 | 36 ± 18 | 0.726 |
| Time since correction, years | 12 ± 17 | 13 ± 19 | 0.792 |
| **Latest echocardiography** | | | |
| Left ventricular systolic fraction ejection | 63 ± 6 | 60 ± 6 | 0.557 |
| Pulmonary artery pressure | 27 ± 9 | 26 ± 8 | 0.618 |
| S' wave cm/s | 9.5 ± 2.3 | 11.4 ± 2.5 | 0.456 |
| Tricuspid annular plane systolic excursion, mm | 17 ± 5 | 21 ± 6 | 0.050 |
| Right ventricular dilation | 46 (36) | 33 (26) | 0.086 |
| Right atrial dilatation | 47 (36) | 31 (25) | 0.033 |
| Right ventricular dysfunction visually | 19 (15) | 8 (6) | 0.085 |
| Valvulopathy | 24 (19) | 33 (26) | 0.166 |
| Pulmonary hypertension | 4 (3) | 5 (4) | 0.734 |
| **Latest exercise testing** | | | |
| Heart beat rate, % predicted | 91 ± 13 | 89 ± 14 | 0.607 |
| MET, % predicted | 97 ± 33 | 95 ± 29 | 0.538 |
| | | | |
| Arrhythmia during follow-up | 48 (37) | 45 (21) | 0.697 |
| Tachyarrhythmia | 47 (36) | 43 (33) | 0.601 |
| Conduction arrhythmia | 11 (9) | 1 (1) | 0.003 |
| Atrial fibrillation, flutter or tachycardia | 41 (32) | 40 (31) | 0.893 |
| Ventricular tachycardia | 5 (7) | 4 (3) | 0.734 |
| Symptomatic | 43 (33) | 40 (31) | 0.689 |
| Medication | 62 (48) | 46 (36) | 0.043 |
| Arrhythmia treated by drugs | 35 (27) | 26 (20) | 0.187 |
| Electrophysiological study | 12 (9) | 7 (5) | 0.233 |
| Pacemaker implantation | 14 (11) | 6 (5) | 0.063 |
| Reoperation | 18 | 6 | 0.010 |

Data are mean ± standard deviation or n (%)