Research proposal for doctoral thesis of Mrs. M. Touray

Long term follow-up of adult patients with anomalous pulmonary venous return

Principal Investigators:

Mariama Touray[a]

Tobias Rutz[a]

Co-investigators

Judith Bouchardy[a]

Nicole Sekarski[b]

[a]Division of Cardiology, Centre hospitalier universitaire vaudois, Lausanne, Switzerland

[b]Division of Paediatric Cardiology, Centre hospitalier universitaire vaudois, Lausanne, Switzerland

Address for correspondence:

Tobias Rutz MD

Service of Cardiology

Centre hospitalier universitaire vaudois

Rue du Bugnon 46

1011 Lausanne

Switzerland

Telephone: +41 79 556 70 33

Fax: +41 21 314 48 02

tobias.rutz@chuv.ch

Introduction

Anomalous pulmonary venous connection (APVC) is a rare congenital heart disease (CHD) defined by pulmonary veins aberrantly attached to the right atrium or a systemic vein. One differentiates partial (PAPVC) and total anomalous pulmonary venous connections (TAPVC) with an incidence of 0.6% in autopsies.(1-4)

The consequence of the anomalous drainage in PAPVC is a left to right shunt with a volume overload of the right heart and of the pulmonary circulation, provoking a dilation of the right cardiac cavities.(5) Presenting symptoms of PAPVC resemble atrial septal defects (ASD) including dyspnea on exercise, diminished exercise capacity and atrial arrhythmia.(6) The clinical symptoms vary depending on the number of anomalous pulmonary and the consecutive amount of shunting. In order to correct this anatomic defect, patients have to undergo surgery consisting in redirecting the anomalous pulmonary vein to the left atrium.

The scimitar syndrome is a particular subtype of PAPVC characterized by an abnormal drainage of all or some right pulmonary veins either above or below the diaphragm into the inferior vena cava.(7) Hypoplasia of the right lung is often associated to this entity. Outcome depends on pulmonary vein obstruction or pulmonary hypertension.(8, 9)

TAPVC is the most severe form of APVC with an incidence of about 0.6 to 1.2 for 10'000 live births.(4) An ASD is necessary for the survival of the newborn. As all veins drain anomaly, symptoms generally develop within the first months of life such as cyanosis and dyspnea requiring surgical correction during early life.(10) Pulmonary venous obstruction is a serious complication and predictor of post-operative mortality.(10-12)

Data on mortality and morbidities of patients with APVC are scarce due to the low incidence.(2, 5) Beside stenosis of a pulmonary vein requiring surgical re-intervention or a percutaneous intervention by cardiac catheterization arrhythmias and loss of sinus rhythm after surgical correction are known complications.(13-17) However, interestingly little is known on outcome of naïve patients without surgical interventions.(5) In addition, indications for surgical correction in patients with PAPVC and only mild left to right shunting are unclear and do not appear in current CHD guidelines.(18)

Aim

The aim of the study is therefore to evaluate outcome of adult patients with APVC included in the Sacher registry and to compare in particular the characteristics of naïve and surgical corrected patients with PAPVC with a special focus on arrhythmias.

Methods

Study design

This will be a retrospective, observational multi-center study. The study will be the doctoral thesis of Mrs. M. Touray, one of the principal investigators.

Patient population

All adult patients diagnosed with TAPVC or PAPVC including scimitar syndrome followed at the institutions participating at the Sacher registry.

Inclusion criteria

- Patients with PAPVC with or without ASD
- Patients with TAPVC
- At least one follow-up report on an echocardiography exam or outpatient visit

Exclusion criteria

- Associated complex congenital heart disease potentially leading to right heart dilatation, pulmonary hypertension such as conotruncal heart defects, complete atrioventricular septum defects, single ventricle physiology etc.

Primary endpoints

- Exercise capacity as documented by cardiopulmonary exercise test or exercise test
- Right and left heart size
- Incidence of arrhythmias

Secondary endpoint

- Hemodynamic parameters such as shunt (Qp:Qs) pre- and at last follow-up, pulmonary artery pressure
- Right and left heart size at diagnosis and at last outcome
- Arrhythmias pre- and post-operatively and their treatment

Further study variables

- Details on surgical correction
- Medical treatment
- Patients' characteristics like gender, age, height, weight etc.

Procedures

- Medical data will be reviewed retrospectively from the electronic and/or paper archiving systems.
- After data collection, patients will be coded for further analysis. Quantative and qualitative analyses will be performed to compare the predefined groups

Statistical analysis

Continuous variables will be indicated in frequencies (%), mean ± standard deviation or median and range, where appropriate. Chi square test will be used to compare categorical variables. The Student *t-test* will be used to compare parametric variables. The statistical significance level will be set at a p-value < 0.05.

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Dr. T. Rutz

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