Case report

Repair of a partial anomalous venous return from the left lung in a patient after atrial septal defect closure

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A partial anomalous pulmonary venous return (PAPVR) is found in around 0.4–0.7% of autopsies [1]. PAPVR is described with a higher incidence in different genetically encoded disease. A typical example is the numerical chromosomal aberrations conditional Turner syndrome with a higher prevalence PAPVR [2]. About 85–90% of these cases are associated with atrial septal defect [3].

A 47-year-old male, ex-smoker with arterial hypertension, bronchial asthma, sleep apnea and migraine in history was admitted to our center with worsening dypsnea. Seven years ago in another center secundum atrial septal defect size of 26 mm was accidentally diagnosed, without evidence of other congenital defects. Pericardial patch closure of the defect was then performed, complicated by postoperative ventricular
fibrillation with subsequent successful cardiopulmonary resuscitation.

The patient was admitted to our center for post-operative gradually over time progressing overall discomfort, dyspnea of NYHA class III and fatigue. On admission, examinations with transthoracic and transesophageal echocardiography were performed. There was no evidence of valvular disease, and kinetics of both ventricles was normal. There was no detectable shunt flow at the atrial septum, and again without evidence of pulmonary vein anomalies. A cardiac catheterization was performed, with no significant findings in the coronary bed. The pulmonary artery pressure was borderline (PAMP 24 mmHg) and thermodilution demonstrated hyperkinetic circulation – cardiac index (CI) 3.7 l/min/m². An overall reduction in exercise tolerance was confirmed during the 6-min walking test. The ventilation-perfusion scintigraphy ruled out a pulmonary embolism. Laboratory examinations excluded anemia, hyperthyroidism and morbus Paget as a cause of the hyperkinetic circulation. Due to the unambiguous interpretation of the results and clinical indicators a CT angiography of the chest was performed. It showed an anomalous left upper pulmonary vein, draining more than half of the left lung into the left brachiocephalic vein (Fig. 1). Subsequently, the anomalous pulmonary vein was selectively catheterized; the widest diameter was 11.5 mm and the left-to-right shunt was quantified as Qp:Qs of the value 1.95:1. Based on these examinations the patient was indicated to redo surgery. A repeat median sternotomy was performed using a cardiopulmonary bypass without cross clamping the aorta and ligation of the anomalous vein close to the left brachiocephalic vein and its anastomosis to the left atrial appendage (Figs. 2 and 3). The operation and postoperative course were uneventful. The postoperative control echocardiography showed normal flow from pulmonary veins into the left atrium without any pressure gradient. Three months later, the patient’s condition is without any dyspnea (NYHA class I) and he is doing very well.

Fig. 1 – Anomalous vein draining more than half of the left lung into the brachiocephalic vein (arrow).

Fig. 2 – Anomalous vein re-directed to left atrial appendage (arrow).

Fig. 3 – Perioperative finding – anomalous vein re-directed to left atrial appendage (arrow).

Discussion

PAVPR often occurs at different genetically encoded disease. A typical example is Turner’s syndrome [2]. Around 85–90% of the PAVPR is associated with atrial septal defect [3]. A partial anomalous return is described in most cases with right-sided pulmonary veins. An anomalous return of right-sided veins may be directed to the superior vena cava, inferior vena cava, vena azygos, right atrium, or coronary sinus. An anomalous return from the left lung is very rare and it is rarely described in literature. It is reported that its incidence is up to ten times lower.

An anomalous return of one of the pulmonary veins is usually not hemodynamically significant and it is usually well tolerated by patients. It becomes important only in case of potentiated left to right shunt with an atrial septal defect. Therefore, when the cardiac atrial septal defects are reported, it becomes necessary to consider the possibility of a potentiation of the left-right shunt by presence of an
anomalous pulmonary venous return, especially because these defects occur very often together.

A very important issue is the timing of the surgery. Hiji et al. recommend operating after a development of early clinical symptoms. Such an approach is chosen due to knowledge of the risks of the surgery redirection, including atrial fibrillation and other arrhythmias and a rare but possible complication which is the stenosis of redirected vein [4]. On the other hand, Elbardissi et al. prefer to operate on asymptomatic patients. The present prevailing opinion is to operate on all asymptomatic patients and on symptomatic patients with Qp:Qs > 1.5, dilatation of the right ventricle, tricuspid regurgitation or pulmonary hypertension [5].

In the particular case described, atrial septal defect was diagnosed and treated at the primary cardiac center, although there was no evidence of an abnormal return of the left pulmonary vein draining the upper lobe of the left lung to the left brachiocephalic vein. Despite the high incidence, the simultaneous occurrence of these two defects is often not taken into consideration. In this case the anomalous venous return was not diagnosed by transesophageal echocardiography, but later, by the chest CT angiography and cardiac catheterization verification. According to some studies diagnostics using echocardiography may in practice fail because they cannot differentiate the outflow of all pulmonary veins to the left atrium [3,4].

In this concrete patient, the re-direction of the anomalous veins to the left atrial appendage was performed. The transesophageal echocardiography was used to assess the gradient of the anastomosis for peri-operative evaluation of vein derivation to the left atrium. It is considered significant in case of more than 4 mmHg. There was no gradient between pulmonary vein and left atrium in our patient.

A proper diagnosis and assessment of the appropriateness and of the timing of cardiac surgery is an elegant method of improving the patient’s hemodynamic status, which prevents a potential heart failure and improves patient’s quality of life. Appropriate preoperative diagnosis is crucial for the quality of life of patients with multiple heart defects.

**Conflict of interest**

There is no conflict of interest.

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**Ethical statement**

The authors declare that the research was done according to ethical standards.

**Informed consent**

We declare that we obtained informed consent from the patient for publication of this case.

**Appendix A. Supplementary data**

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.crvasa.2014.08.003](https://doi.org/10.1016/j.crvasa.2014.08.003).

**REFERENCES**


