

## Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"  
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

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#### **CASE SUMMARY**

The Authors presented a case of 22-year-old man after pulmonary valve valvulotomy with ASD sinus venosus type. The surgery was performed in 1991. The patient presents at the time the symptoms of easy fatigue, dyspnea, decrease in exercise tolerance. Right Heart Catheterization showed correct values of pulmonary circulation pressures. In echo, CT and MRI the ASD of maximum size of 14 mm was shown. Additional moderate regurgitation through pulmonary valve – netto 59 ml/cycle with maximal speed 1.4 m/s was shown. In spirometry test peak oxygen uptake was 23,7 ml/kg/min and maximal workload was 13,1 METs. Bodyplethysmography showed normal ventilation and respiratory resistance.

#### **DISCUSSION**

Atrial Septal Defects are often combined with other malformations, as abnormal venous return and mitral stenosis (Lutembackers syndrome).<sup>1</sup> Different surgical management techniques for pulmonary stenosis all lead to a good and long-lasting relief of elevated right ventricular pressure, but this is achieved at the cost of pulmonary regurgitation in a third of the patients. Patients operated upon with a transannular patch are at increased risk for reoperation. Especially right atrial and ventricular dilatation and paradoxical septal motion are predictive for pulmonary regurgitation with need of reoperation. Supraventricular arrhythmias disappears after reoperation. Pulmonary regurgitation may account for some impairment in clinical condition during time in these patients.<sup>2</sup> Sinus venosus atrial septal defect (SVASD) encompasses approximately 4% to 11% of atrial septal defects (ASDs). The typical malformation is an interatrial communication caused by a deficiency of the common wall between the superior vena cava (SVC) and the right-sided pulmonary veins. SVASD is commonly associated with anomalous pulmonary venous connection (APVC) of some or all of the pulmonary veins, which produces additional left-to-right shunting. The basic principle of repair is redirection of the APVC through the interatrial communication into the left atrium. In contrast to operative repair of secundum ASD, the surgical approach for SVASD is more complex and carries the risk of stenosis of the SVC or pulmonary veins, residual shunting, and sinoatrial node dysfunction (SND).<sup>3</sup> Surgical repair has low mortality (1% in

patients without significant co-morbidity) and good long-term outcome (normal life expectancy and low long-term morbidity) when performed early (childhood, adolescence) and in the absence of pulmonary hypertension. However, mortality may be higher in the elderly and in patients with co-morbidities. Outcome is best with repair at age <25 years. ASD closure after the age of 40 years appears not to affect the frequency of arrhythmia development during follow-up. However, patients benefit from closure at any age with regard to morbidity (exercise capacity, shortness of breath, right heart failure), particularly when it can be done by catheter intervention. Basing upon ESC guidelines Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms.<sup>4</sup>

## EXPERT'S OPINION

Pulmonary regurgitation may account for some impairment in clinical condition during time in patients after pulmonary valve valvulotomy. Despite the complexity of the lesion, repair of SVASD with associated APVC is associated with low morbidity and mortality even in patients older than 40 years. Severe complications are rare, and development of SND and the need for pacemaker implantation are uncommon. Functional improvement is expected irrespective of age at repair, but postoperative atrial fibrillation appears to be related to older age at operation. SVASD repair achieves survival rates similar to those of a matched population, and although repair is suggested as early as possible, it should be considered whenever repair may impact survival or symptoms.

## CONCLUSION

Basing upon the ESC guidelines the patient should undergo ASD closure procedure if RV volume overload symptoms are present. Pulmonary valve regurgitation seems not to be the main source of his symptoms. Although the problem should be discussed with cardiac surgeon performing the ASD closure.

## REFERENCES

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