**Article**

"Development of the European Network in Orphan Cardiovascular Diseases"

„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

**Title: Patients with interatrial communications**

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Patients with interatrial communications (atrial septal defect and patent foramen ovale) form the largest group of adults with congenital heart defects. Patent foramen ovale (PFO) is the most common interatrial communication that is found in about 26% (17-35%) of the population, whereas atrial septal defect (ASD) makes up about 10% of all congenital heart defects at birth and as much as 30-40% of all congenital heart defects in adults over 40 years of age.

***Atrial septal defect***

Atrial septal defects arise from an abnormal development of the endocardial cushions or the second septum of the fetal heart, leading to preservation of the communication between the atria.

There are four types of atrial septal defects depending on their location in the heart:

* Atrial septal defect ostium secundum type (ASD ostium secundum, ASD II ),
* Atrial septal defect ostium primum type
* Sinus venosus atrial septal defect
* Coronary sinus atrial septal defect

Because of long-term asymptomatic course atrial septal defects are frequently detected in adults. The commonest presenting symptom is exertional dyspnea, reduced physical capacity, paroxysmal heart palpitations. There is sometimes increased predilection to respiratory infections, peripheral edema, atypical chest pain and syncope. The atrial septal defect may be an incidental finding in diagnostic tests for other reasons. The clinical symptoms develop slowly and for this reason the patients remain asymptomatic until the first decade of life. On the other side, the percentage of asymptomatic adults over 40 years of age is small.

The reasons for progression of the clinical symptoms of ASD include age-dependent decrease in left ventricular compliance enhancing left-to-right shunting, pulmonary hypertension and atrial fibrillation which frequently leads to right ventricular failure.

Hemodynamically significant ASDs should be closed in adults if pulmonary vascular resistance (PVR) does not exceed 6-8 Wood units. PVR which is significantly reduced after oxygen, tolazoline or nitric oxide is also an indication for ASD closure.

There is controversy regarding the usefulness of ASD closure in patients with insignificant left-to-right shunts, absent pulmonary hypertension or in adults over 40-60 years of age. The search for potential indicators that would help establish indications for the intervention continues in patients with borderline and asymptomatic ASDs.

The closure of an isolated ASD in adults is considered a safe procedure with mortality rates of 0-1.3%. Percutaneous closure of ASD II was first attempted by King and Mills in 1976. Since then both the techniques and the devices have been improved. The outcomes of ASD closure are promising and percutaneous transcatheter correction is the treatment of choice in patients with ASD II according to the European Society of Cardiology. The procedure has been found safe and effective in a vast majority of patients.

Atrial fibrillation is the most frequent atrial tachyarrhythmia in patients with ASD. Atrial flutter or supraventricular arrhythmia is rare. The incidence of atrial fibrillation increases with age. In the literature there are only single reports on arrhythmias complicating percutaneous transcatheter correction of ASDs or clinical course after the procedure.

***Patent foramen ovale***

In 1877 Cohnheim, German anatomical pathologist who performed a post mortem study of a young female hypothesized that brain stroke had been caused by paradoxical (crossed) embolism. For many years the presence of patent foramen ovale (PFO) has been associated with increased risk of crossed embolism leading to cryptogenic brain stroke. This relationship has been found mainly in young patients with cryptogenic strokes, but it is suspected also in older subjects.

In Poland about 60 000 brain strokes are detected annually, of which nearly 30% (18 000) are cryptogenic. Assuming that every third cryptogenic stroke can be associated with PFO, it is about 6000 strokes per year, posing a serious clinical, economic and social problem. There is a strong correlation between the size of PFO and the severity of neurological symptoms in patients with transient ischemic attacks.

There is no standard approach to patients with PFO and ischemic stroke, regarding both pharmacological treatment and surgical/percutaneous correction. Although there are no clear indications for PFO closure it appears reasonable to perform the procedure in patients with recurrent cryptogenic embolism, especially when the PFO coexists with severe or moderate contrast passage to the left atrium on echocardiography during Valsalva maneuver, and/or atrial septal aneurysm. The size of PFO and the presence of Eustachian valve appear to be important. There are only single reports of the relationship between the morphology of the foramen ovale and the risk of crossed embolism.

However, the approach to PFO patients still raises controversy regarding not only the technique of PFO correction, but also whether such patients should be treated at all. It is still unclear whether each PFO patient should receive treatment, or whether treatment should be instituted after the first transient ischemic attack, or whether we should wait until more cerebrovascular events occur.

Patent foramen ovale is one of the most common cardiovascular anomalies in an adult population (1 in every 4 people), therefore it is an obvious mistake to treat every patient with PFO. The detection of a large significant PFO raises doubt because of difficulties in the interpretation of the Valsalva maneuver. The Valsalva maneuver during transesophageal echocardiography may be ineffective as it requires patient cooperation after he/she has been properly instructed. Furthermore, the result may be incorrectly interpreted, thus distorting the significance of PFO.

An accurate morphological and functional assessment of PFO is of practical value as it is possible to identify patients with an increased risk of crossed embolism.

The available evidence shows that PFO closure should not be an individualized rather than routine procedure in patients with cryptogenic strokes. Every patient should undergo neurological diagnostic procedures with imaging that would confirm brain ischemia and then assigned to pharmacological or interventional therapy.

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Author’s signature\*

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