Medical Expertise

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

EXPERT: Prof. Piotr Podolec, MD, PhD, cardiologist

Affiliation: *Department of Heart and Vascular Diseases, John Paul II Hospital, Cracow, Poland*

CASE SUMMARY

The authors presented a case of a 42-year-old patient with aortic aneurysm and a complex congenital heart disease: ventricular septal defect and patent ductus arteriosus. At the age of 2 he underwent PDA closure and pulmonary artery banding (Damman-Muller procedure). At the age of 18 detailed cardiologic evaluation was required due to dyspnea and cyanosis. Right heart catheterization showed significant left-to-right shunt and increased pressure gradient in RVOT. Total surgical correction was performed: VSD was closed and pulmonary artery was extended by insertion of a cylindrical patch.

In 2010 cardiac angio-CT showed enlargement of the aortic annulus (46 mm), aortic bulb (54 mm), the aortic arch was 38 mm wide. No PDA flow, no leakage through the interventricular septum, no stenosis of the pulmonary trunk and no pathology in the coronary arteries were detected. The examination disclosed a vascular anomaly: double superior vena cava with the left one terminating in the coronary sinus. A check-up angio-CT performed in January 2014 showed: right aortic arch, further widening of the ascending aorta, the dimensions of the aortic arch and the descending aorta were within normal limits.

The patient underwent further cardiac evaluation. He suffered from decrease in exercise tolerance. Co-existing diseases were: mild mental retardation, obesity, arterial hypertension, hepatitis C, history of DVT of the left lower limb. On physical examination he was in good general state, BP was normal, heart rate was 70 bpm, diastolic murmur in the 2nd left intercostal space was noticed, no cyanosis was observed. Furthermore, obesity, enlarged liver, enlarged left lower limb circumference. ECG showed regular sinus rhythm 70 bpm, normal heart axis, right bundle branch block. Because of elevated glucose level, oral glucose tolerance test was performed and impaired fasting glycaemia was diagnosed. The transthoracic echocardiography showed enlarged right atrium and right ventricule, preserved left ventricular ejection fraction (EF 65 %), enlarged ascending aortic: aortic annulus - 31 mm aortic bulb – 55 mm, ascending aorta – 53 mm, aortic arch 29 mm, moderate aortic regurgitation, turbulent flow in RVOT, pulmonary valve gradient 25/12 mmHg. No signs of leakage through IVS was noticed. Cardiac angio-CT showed ascending aortic aneurysm without dissection. The aortic dimensions were comparable to the previous results. Moreover,

coronary arteries were without atherosclerotic changes, there was a myocardial bridge narrowing the left anterior descending artery, there was no PDA flow or leakage through the interventricular septum, no stenosis of the pulmonary trunk, the aortic valve was tricuspid. 24-hour Holter ECG monitoring showed no significant abnormalities.

DISCUSSION

Ventricular septal defect is the second most common congenital heart defect (30-40%), the first being the bicuspid aortic valve. Patent ductus arteriosus (PDA) often coexists with other congenital heart abnormalities. Nowadays the Damman-Muller procedure (banding of the pulmonary artery) is a rarely performed palliative operation - it's aim is to reduce excessive pulmonary blood flow (in the case presented above caused by the VSD - related left-to-right shunt) in order to prevent development of arterial pulmonary hypertension.

In ascending aortic aneurysm surgery is indicated when aorta's dimension is above 5-5.5 cm. Another argument for surgery is increasing the size of the aneurysm >2-4 mm per year.

EXPERT'S OPINION

In the presented case due to dimensions of the aneurysm and it's progressive enlargement surgical treatment is recommended. It appears that replacement of the aorta along with own aortic valve sparing would be the optimal solution (one have to consider possible non-compliance during VKA therapy). Because of a history of deep vein thrombosis, ventilation/perfusion scintigraphy should be performed to exclude pulmonary embolism. Moreover, additional diagnostics of coagulation system is indicated. Right heart catheterization should also be considered.

CONCLUSION

Surgical management is recommended. The patient should be evaluated by a cardiac surgeon.

REFERENCES

1. Podolec P., Gackowski A. Tętniaki aorty. In: Podolec P, Tracz W, Hoffman P, editors. Echokardiografia praktyczna tom II. Kraków, Medycyna Praktyczna; 2005. p.262
2. Westaby S, Bertoni GB. Fifty years of thoracic aortic surgery: lessons learned and future directions. Ann Thorac Surg 2007; 83: S832-S834
3. ESC Guidelines for the management of grown-up congenital heart disease. European Heart Journal (2010) 31, 2915–2957

John Paul II Hospital in Kraków
Jagiellonian University, Institute of Cardiology
80 Prądnicka Str., 31-202 Kraków;
tel. +48 (12) 614 33 99; 614 34 88; fax. +48 (12) 614 34 88
e-mail: rarediseases@szpitaljp2.krakow.pl
www.crcd.eu