Medical Expertise

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

EXPERT: Krzysztof Bederski MD, thoracic surgeon

Affiliation: *Department of Thoracic Surgery, John Paul II Hospital, Krakow, Poland*

CASE SUMMARY

Case of 52 year old patient with aneurysm of ascending aorta, congenital heart disease - composite aortic valve disease - moderate aortic stenosis with z severe aortic insufficiency and persitent ductus arteriosus Botalli. Additional the patient has paroxysmal atrial fibrillation, and hypertention.
In echocardiography performed during years 2009 – 2014 extending of ascending aorta from 46 mm till50 mm, increase of aortic gradient (from 32/20 mmHg till 41/23 mmHg), enlargment of left ventricle (from 54/35 mm till 60/40 mm) were observed.
Actually in echocardiography ejection fraction is 60%, dimension of left ventricle: 60/40 mm, ascending aorta – 50 mm, arch of aorta 30 mm, separation of aortic valve leaflets 8 mm, aortic gradient 41/23 mmHg, moderate/severe aortic insufficiency – PHT 300 and suspicion of biscuspid aortic valve.

In angio CT (2013) aneurysm of ascending aorta was observed and dimesions were measured: aoritc bulb 44 mm, ascending aorta 51 mm, aortich arch in truncus brachioencephalicus - 37 mm, aoritc arch in left subclavian artery – 27 mm, descending aorta in proximal part - 33 mm, descending aorta in distal part - 25 mm. Also calcifications of aortic valve leaflets were observed. The persistent ductus arteriosus Botalli was seen and longitudinal dimension was 26 mm, diameter about 4-5

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

mm, vestigial flow to pulmonic trunc.
In coronarography there was not atherosclerosis in coronary arteries and dilatation of ascending aorta to 48 mm, calcification in aortic valve, moderate/severe aortic valve insufficiency were observed. Coronarography was complicated by pseudoaneurysm (treated by operation).

DISCUSSION

According to actual ESC guidelines for the management of grown – up congenital heart disease (2010) PDA should be closed in patients with signs of LV volume overload (class I c) and in patient with pulmonary arterial hypertension but pulmonary artery pressure < 2/3 of systemic pressure or pulmonary vascular resistance < 2/3 of systemic vascular resistance (class I c) Device closure is the method of choice were technically suitable (class I c). Surgery is reserved for patients with a duct too large or with unsuitable anatomy.

Additionally according to ESC guidelines on the management of valvular heart disease (2012) the patient has class I c indication for surgery in aortic root disease (whatever the severity of AR surgery is indicated in patient who have aortic root disease with maximal ascending aortic diameter =>50 mm).

EXPERT'S OPINION

Right heart catheterization would in this patient be recommended for the evaluation of hemodynamic parameters and the evaluation of shunt through the PDA. In this patient closure of PDA seems to be necessary. Both the PDA closure and surgical correction of aortic valve disease at the same time should be considered. Final decision should be taken after results of cardiac catheterization and quantification of AR severity in echocardiography.

REFERENCES

ESC guidelines on the management of valvular heart disease (2012).
ESC guidelines for the management of grown – up congenital heart disease (2010)

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