

Pregnancy in patient with Turner syndrome, after surgery for coarctation of the aorta, with bicuspid aortic valve and ascending aortic aneurysm (RCD code: VII-I-1B.6)

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Abstract

A 30-year-old woman with Turner Syndrome was admitted to our centre in 12th week of her first pregnancy with a history of surgical correction of coarctation of aorta. She was asymptomatic with good general condition. Transthoracic echocardiography revealed a bicuspid aortic valve with moderate regurgitation and an ascending aorta aneurysm. In angio-MR the narrowing of the isthmus of the aorta over 50% was seen with peak systolic gradient found in echocardiography approximately 41 mmHg. She was prescribed metoprolol and was closely monitored, the course of pregnancy was uneventful. In 24th week of pregnancy steroids were administered as a preparation for earlier delivery. She delivered in 34th week of pregnancy a healthy baby weighting 2100 grams. Caesarean section was performed in the cardiothoracic surgery ward and after delivery she underwent percutaneous angioplasty of coarctation of aorta with stent implantation. Six weeks after labour she underwent the Bentall de Bono procedure with artificial aortic valve implantation. In six-month observation patient was in good general condition, echocardiography showed normal ascending aorta diameter. JRCd 2013; 1 (4): 150–154

Key words: pregnancy in patient with Turner syndrome, coarctation of aorta

Case presentation

A 30-year-old Caucasian woman with Turner Syndrome (karyotype 80% 45,X0; 15% 46,XX; 5% 47,XXX) was admitted to the hospital in 12th week of pregnancy. She has a coarctation of aorta surgically corrected at the age of 2 years ('end-to-end' procedure), bicuspid aortic valve and ascending aorta aneurysm. Patient's height was 139 cm, weight 46 kg, body mass index 23,5 kg/m², body surface area 1,3 m². She wasn't previously under systematic cardiologic supervision. The diagnosis of the aortic aneurysm was first made on the ground of echocardiography examination in the 4th week of pregnancy. Patient was informed about the high risk connected with pregnancy.

She was asymptomatic with good general condition.

Laboratory test results showed no abnormalities.

In physical examination diastolic murmur over the base of the heart was heard.

The oxygen saturation of arterial blood was 94% at rest.

In electrocardiogram (ECG) at rest sinus rhythm with heart rate 80/bmp was seen.

The 24-hour ECG Holter monitoring didn't show significant arrhythmias or conduction disturbances.

The blood pressure in 24-hour monitoring was normal. The blood pressure measured on the left upper extremity was approximately 24 mmHg lower than on the right one, blood pressure on lower extremities was equal to the value measured on the left arm.

Conflict of interest: none declared.

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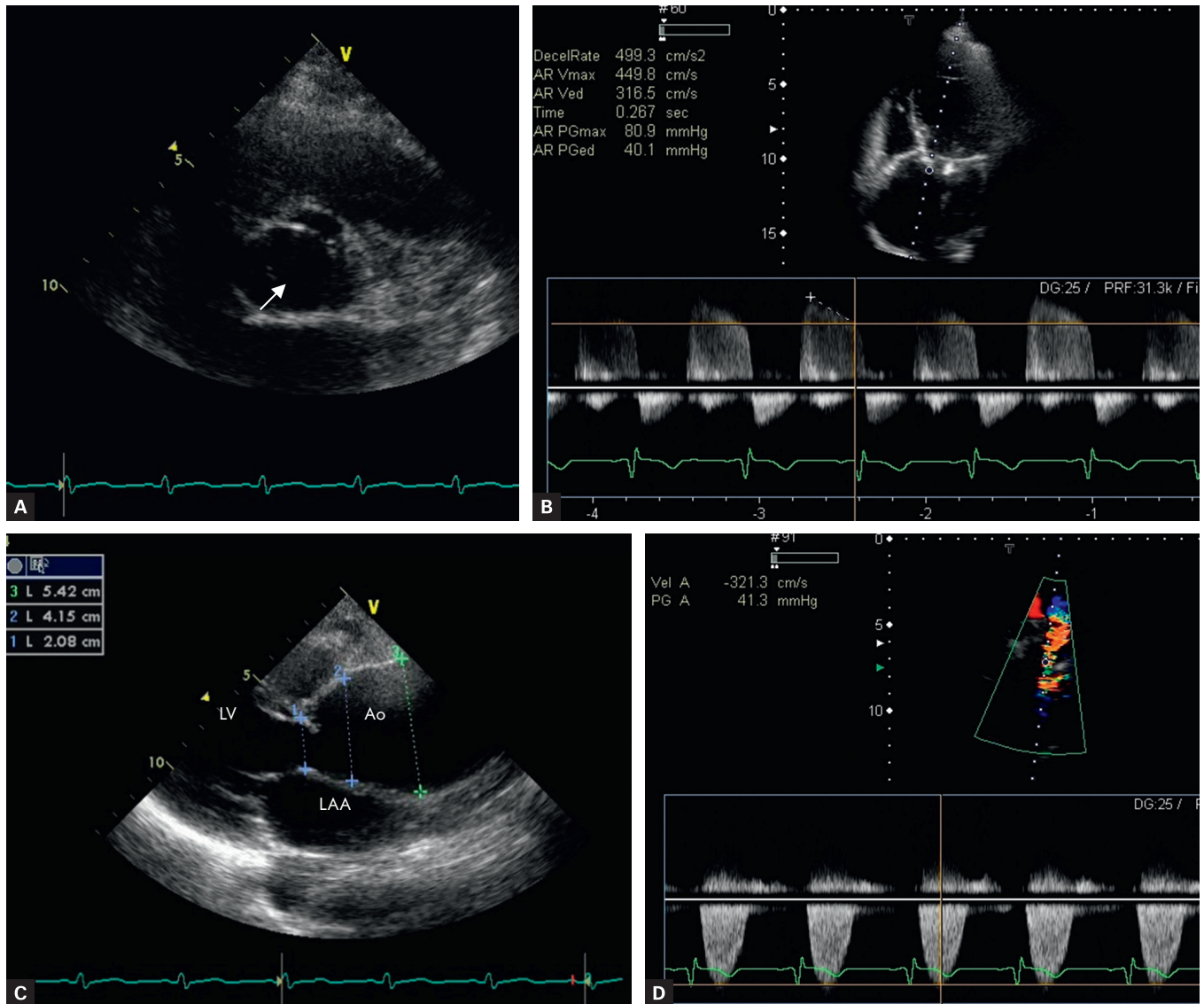


Figure 1. Echocardiography: A. Parasternal short axis view. Bicuspid aortic valve (white arrow); B. Continuous wave Doppler ultrasound showed moderate aortic valve regurgitation; C. Parasternal long-axis view. The ascending aortic diameter was extremely enlarged 54 mm; aortic index 39 mm/m²; D. Suprasternal view. Systolic gradient of coarctation of the aorta 41 mmHg

Transthoracic echocardiography revealed normal right and left ventricles size and function, normal both atria diameter, bicuspid aortic valve with moderate regurgitation (Figure 1.A, B). The ascending aorta was dilated to 54 mm, aortic index 39 mm/m² (Figure 1C). The peak systolic gradient of coarctation of aorta was approximately 41 mmHg (Figure 1D). The Doppler ultrasound imaging showed retrograde vertebral flow on the left side and normal diameter of the left subclavian artery. Normal flow in the right vertebral artery, both-side carotid and femoral arteries was found.

In *angio-MR* the narrowing of the aortic isthmus over 50% was seen (Figure 2). The ascending aorta's diameter was 5,5x5,5 cm, than narrowing to 2,7x2,8 cm.

Review of literature

In most cases of the patients with Turner Syndrome there is an issue of primary hypertension, usually connected with cardiovascular or kidney diseases, including coarctation of the aorta. According to the sources, the prevalence of the aortic root dilatation is estimated from 8,8% to 42% in patients with gonadal dysgenesis [1,2]. In the analysis of 85 patients with Turner Syndrome after aortic dissection, in 89% of them at least one established risk factor of aortic rupture was present, including coarctation of aorta, bicuspid aortic valve and hypertension [3]. Most of the patients with Turner Syndrome are sterile due to gonadal dysfunction. Oocyte donation is usually the only possibility to become pregnant for this women [5,6]. Nevertheless, cases of patients with Turner mosaicism who successfully become pregnant and carried their pregnancies to term are described in literature [7].

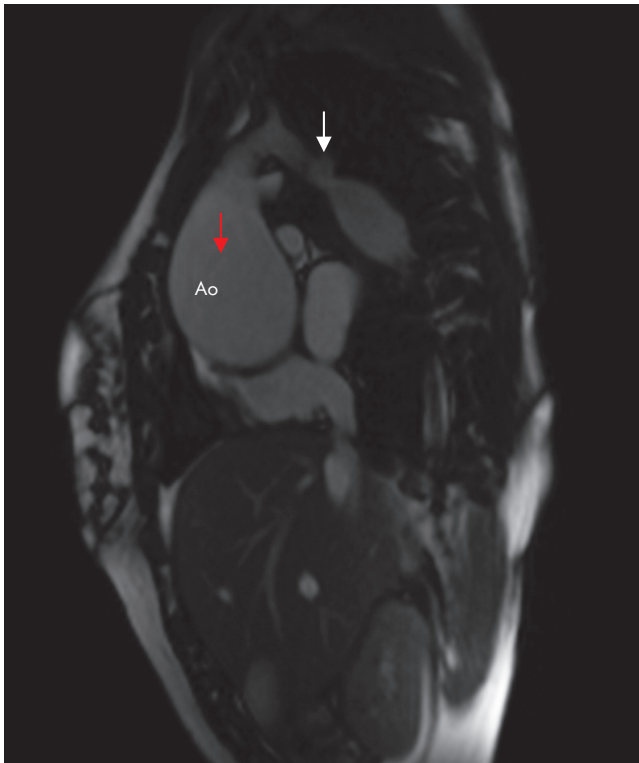


Figure 2. Cardiac magnetic resonance showed ascending aorta dilatation (red arrow) and narrowing of the isthmus of the aorta above 50% (white arrow)

There is a risk of cardiovascular complications of pregnancy in patients with Turner Syndrome. In several studies of this population, increased risk of aortic dissection was suggested, especially if risk factors for aortic rupture were present – such as coarctation of aorta, bicuspid aortic valve and hypertension. The maternal mortality associated with aortic complications during pregnancy reaches 11% and is mainly attributed to type A dissection. Also risk of (pre) eclampsia is elevated. Treatment of hypertension, especially during pregnancy, is crucial [8,9,10].

The 2011 European Society of Cardiology Guidelines of management of cardiovascular diseases during pregnancy include patients with Turner Syndrome and aortic aneurysm to WHO risk Class IV (the modified World Health Organization risk classification) [11].

Patients with aortic dilatation >50 mm associated with bicuspid aortic valve and severe coarctation are advised not to get pregnant, however if they become pregnant and will not consider termination, review ever month or twice a month is necessary.

In this particular case, due to small height, patient's aortic diameters must be recount in relation to body surface area. The aortic diameter index 27 mm/m² is connected with an increased risk of aortic dissection and surgery intervention should be consider before pregnancy [11].

Follow up with echocardiographic imaging should be repeated every 4 to 8 weeks in pregnant patients with ascending aorta dilatation. Administration of beta blockers during pregnancy is advised [11].

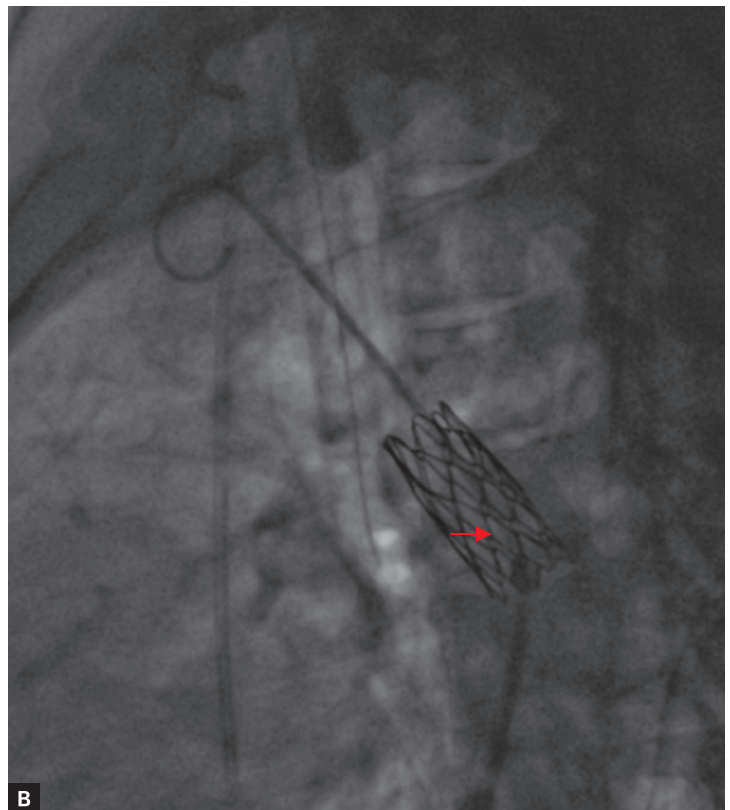
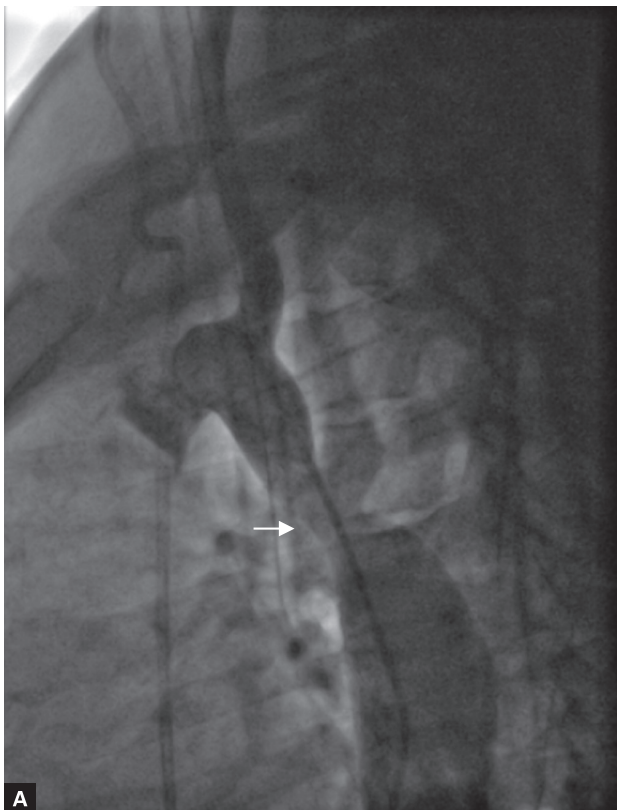


Figure 3. Aortography. A. Coarctation of the aorta (white arrow); B. Percutaneous baloon angioplasty of coartation of aorta with stent implantation (red arrow)

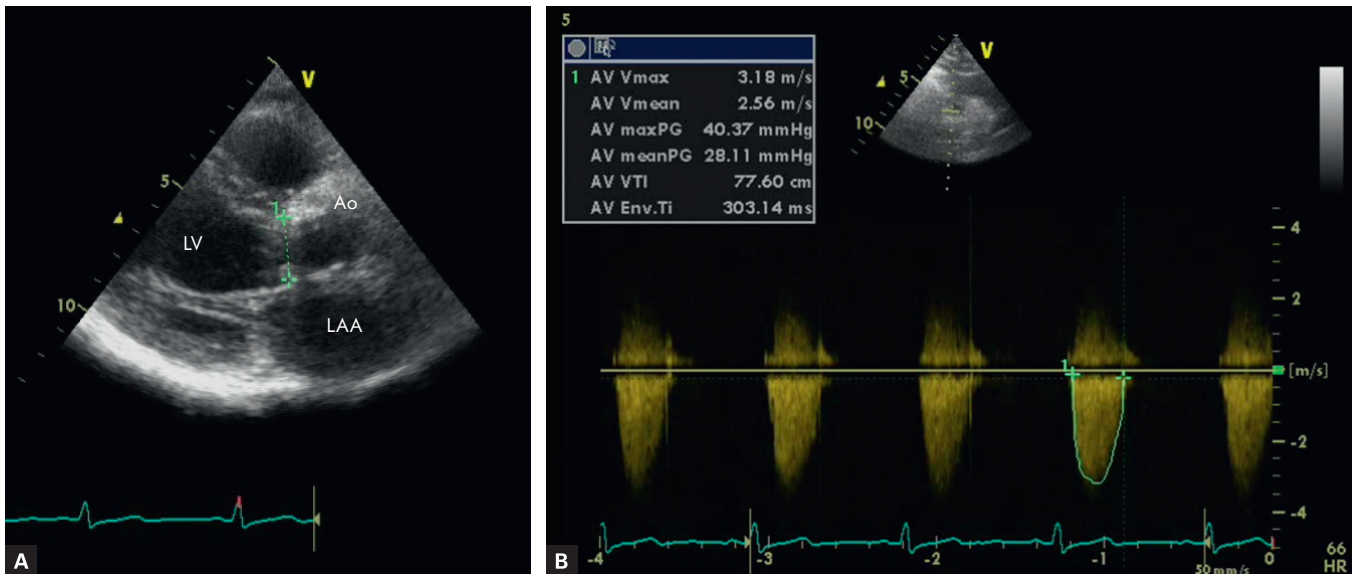


Figure 4. Echocardiography after Bentall de Bono procedure A. Parasternal long axis view. – normal aortic diameter; B. Suprasternal view. Systolic gradient of coarctation of the aorta 40 mmHg

In case of progressive aortic dilatation or aortic rupture during pregnancy, before the labour is possible, aortic surgery with fetus in utero should be considered. When fetus is viable, caesarean section followed directly by aortic surgery is recommended. Delivery should be performed in a hospital with cardiothoracic surgery ward and neonatal intensive care facilities [11].

Patient management and follow-up

In medical treatment Metoprolol 75 mg/daily was administered. The patient was closely clinically monitored with echocardiographic evaluation every two weeks until delivery. She was clinically stable with normal blood pressure and no changes in left ventricle size and aortic diameter in repeated echocardiography.

After gynecologic consultation in 24th week of pregnancy steroids were administered as a preparation for earlier delivery. The patient gave birth to a healthy baby with normal karyotype at 34th week of pregnancy, Apgar score was 9, weight at birth was 2100 g. According to the ESC guidelines cardiologists together with the gynecologist decided to perform a *Cesarean section*, which was performed in the cardiothoracic surgery ward and after delivery, patient underwent *percutaneous angioplasty of coarctation of aorta with stent implantation* (Figure 3). After successful operation gradient in the aortic isthmus was reduced from 20 mmHg to 0. Patient was stable, aortic aneurysm diameter was stationary. Six weeks after labour patient underwent *the Bentall de Bono procedure* with artificial aortic valve implantation. In six-month observation patient was in good general condition, echocardiography showed normal ascending aorta diameter (Figure 4). This case was discussed during 2nd Symposium on Rare Cardiovascular Diseases – ESC Munich 2012 [4].

Algorithm

Before pregnancy: Systemic hypertension associated with high risk of aortic dissection. In echocardiographic imaging: bicuspid aortic valve, coarctation of aorta, ascending aorta aneurysm –high risk factors of aortic dissection. The aortic diameter index 27 mm/m^2 – prophylactic aortic surgery before pregnancy should be considered.

During pregnancy: Patient in WHO risk Class IV – with systemic hypertension, severe coarctation, bicuspid aortic valve and the aneurysm of the aorta. Bimonthly clinical and echocardiographic monitoring with blood pressure measurement is recommended. Beta blocker and hypertension therapy should be administered.

Delivery: Caesarean section in hospital with cardiothoracic surgery ward and neonatal intensive care facilities should be performed.

In case of progressive aortic dilatation or aortic rupture during pregnancy, before the labour is possible, aortic surgery with fetus in utero should be considered. When fetus is viable, caesarean section followed directly by aortic surgery is recommended [11].

After pregnancy: Patient should undergo *The Bentall de Bono procedure* with artificial aortic valve implantation.

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All the study have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki. Patient gave her informed consent prior to inclusion in the study.