Aortic root aneurysm in a patient with Marfan syndrome and D-transposition of the great arteries

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We present a case of a 13-year-old boy with Marfan syndrome and an aortic root aneurysm. In medical history, the child was diagnosed at birth with D-transposition of the great arteries (D-TGA) and operated on with arterial switch operation (ASO) with the Le Compte maneuver. The diagnosis of Marfan syndrome was established at the age of 3 years. Follow-up transthoracic echocardiography (TTE) showed a progressive life-threatening aortic root aneurysm requiring cardiac surgery. On cardiac evaluation, the patient was in good condition. Vital signs showed a regular heart rate of 70/min, blood pressure of 110/70 mm Hg, and normal oxygen saturation (SaO_2) >95%. The Marfan phenotype was found on physical examination with a tall and slender silhouette (body weight 53 kg, height 1.8 m, bovine serum albumin [BSA], 1.72 m^2), scoliosis, pectus deformity, arachnodactyly and nearsightedness.

Transthoracic echocardiography (TTE) and magnetic resonance imaging showed left ventricular dilation (end-diastolic diameter 60 mm, z score +2.4, end-systolic diameter 43 mm, z score +3.0), decreased contractility (ejection fraction [EF], 55%), significant aortic annulus dilatation (37 mm, z score +6.9), aortic root aneurysm (51 mm, z score +6.3 measured from inner edge to inner edge at the widest diameter in diastole) and severe aortic valvar regurgitation (Figure 1A–C; Supplementary material, *Video S1–S3*). The patient was qualified for the Bentall procedure in cross-clamp circulation. The pulmonary trunk was in front of the aorta following the Le Compte maneuver and required resection to get access to the aneurysm. Aortic root replacement was performed with a mechanical prosthetic valve (29 mm SJM masters HP series Valved Graft) followed by coronary arteries re-implantation. Finally, a 22-mm valved prosthesis was implanted at the position of the pulmonary trunk with an additional 16 mm prosthesis into the pulmonary confluence creating a letter "T". Hemostasis was a severe problem, and the patient required blood, fresh frozen plasma, and platelets transfusions as well as a TachoSil Fibrin Sealant Patch. The postoperative period was complicated by low cardiac output and poor myocardial contractility. Therefore, mechanical circulatory support was established with extracorporeal membrane oxygenation (ECMO) via the femoral vein and aortic arch cannulas. The patient was unstable despite multidrug therapy with adrenaline, dopamine, milrinone, and levosimendan infusions.

Coronary angiography revealed features of distal embolization of the right coronary artery (Figure 1D; Supplementary material, *Video S4–S6*). Pleural and pericardial bleeding required chest revision, with ECMO cessation and a simultaneous Nuss procedure, with titanium steel bar implantation correcting chest deformity. Right heart catheterization was performed and a 12 mm × 39 mm BeGraft balloon expandable *covered stent* (*Bentley* InnoMed, Hechingen, Germany) with low foreshortening and high radial force was deployed into the stenotic right pulmonary artery. The stent was redilated with a 16 mm Tyshak balloon catheter, with simultaneous left pulmonary artery angioplasty (Figure 1E; Supplementary material, *Video S7–S9*). Within

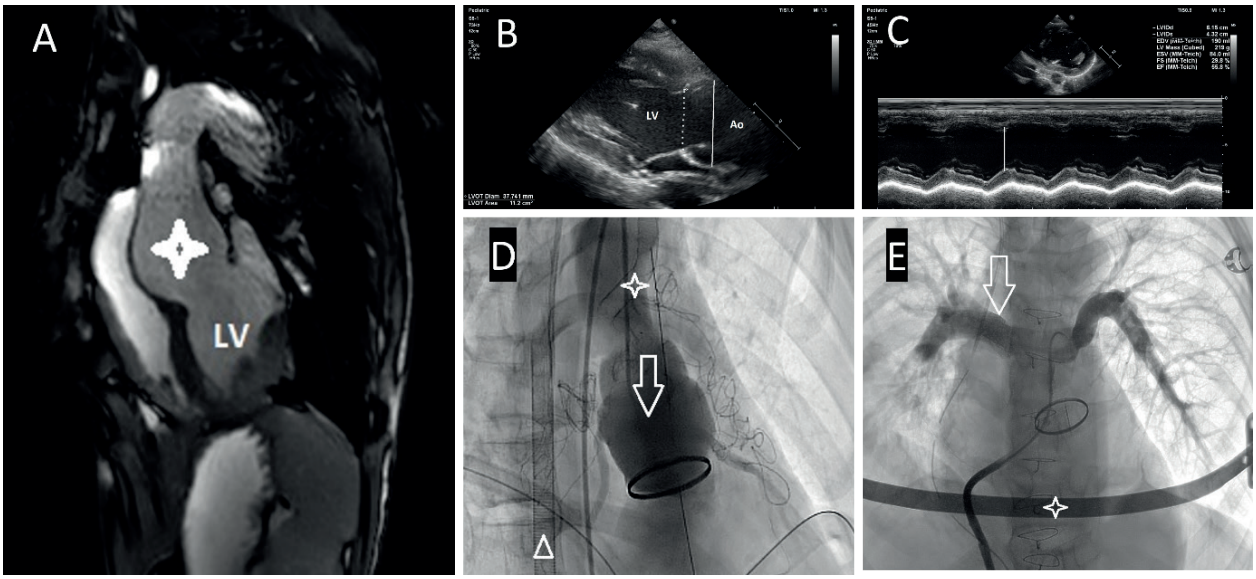

Figure 1. A. MRI. Lateral view showing dilated left ventricle and aortic root aneurysm (white star). **B.** TTE. Parasternal long axis view. Dilated aortic valve 37 mm (dotted line) and aortic root aneurysm 51 mm (white line). **C.** TTE. M-mode in parasternal long axis view of the left ventricle showing dilated end-diastolic and end-systolic (white line) diameter with decreased ejection fraction of 55%. **D.** Aortography in RAO 30° projection. Extracorporeal circulation with aortic (white star) and venous (white triangle) cannulas. Aortic prosthesis (white arrow) with artificial aortic valve (black eclipse). **E.** Pulmonary trunk angiography in cranial view (38°) showing right pulmonary artery with a covered stent (white arrow) and left pulmonary artery following balloon angioplasty. A steel bar (white star) implanted during Nuss procedure due to chest deformity

Abbreviations: MRI, magnetic resonance imaging; RAO, right anterior oblique; TTE, transthoracic echocardiography

two weeks TTE showed improvement with left ventricular ejection fraction (EF, 42%) and right ventricular fractional area change (40%). Chronic respiratory failure required a temporary tracheotomy. The patient was discharged home in good condition with multidrug therapy (warfarin, angiotensin-converting-enzyme inhibitor, and verospiron).

Conclusion: Patients with congenital heart defects require lifelong follow-up and reoperation of significant residual or newly emerging lesions, especially with coexisting Marfan syndrome, which predisposes to progressive aortic root dilation requiring an extensive range of operations with high risk of postoperative complications [1–5].

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/kardiologia_polska.

Article information

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