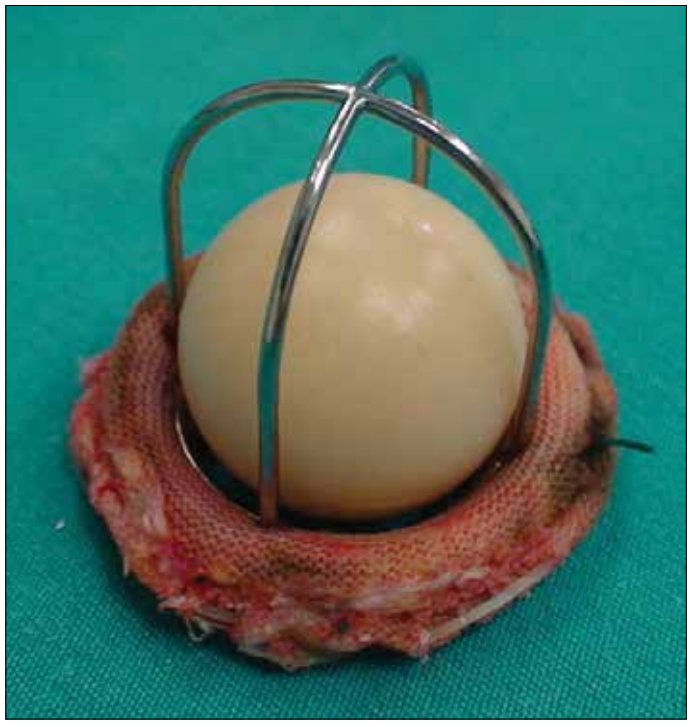

Figure 1. Removed Starr-Edwards mitral valve had no growth of pannus, structural integrity loss and also had no lipid infiltration on the surface of the silastic ball

Edwards laboratories had developed different series of ball valves during next 12 years (1). However, some defects due to the design of the valve such as unacceptable transvalvular gradient in smaller sizes, absence of central flow causing higher transvalvular gradients especially in aortic position and thromboembolic complications associated with the strut clothes had never been solved (2). The incidence of complications related to the Starr-Edwards valve, especially thromboembolism was higher compared with the bileaflet valves (3-5). Regarding these data, even in cases with functioning valves surgeons were tended to replace it with updated versions (6). However, except strut cloth, valve related complications were rare. The loss of structural integrity has only been reported only 2 times for mitral position (2). Recently published results of fifteen years follow-up suggest Starr-Edwards caged ball as a good choice for mitral position (7). Furthermore, several impressive durability case reports are available in the literature. Also Gögje et al. (8) published their thirty year experience with Starr-Edwards prosthesis and they recommend that caged balls after 20 -year durability should not be removed except patients who require additional cardiac operation for other indications.

Conclusion

In our case, patient was suffering from severe tricuspid regurgitation and he did not respond to the medications so operation decision was made. According to literature, we have performed tricuspid valve annuloplasty and mitral valve re-replacement with removing caged ball not to jeopardize patient`s rest life.

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Treatment of aortic valve stenosis and gastrointestinal bleeding by transcatheter aortic valve implantation in Heyde syndrome

Heyde sendromunda transkateter aort kapak implantasyonu ile aort stenozu ve gastrointestinal kanamanın tedavisi

Introduction

Transcatheter aortic valve implantation (TAVI) has recently emerged as an effective therapeutic option for patients with symptomatic, degenerative aortic stenosis (AS) and absolute or relative contraindications to surgical aortic valve replacement. The characterization of Heyde syndrome now refers to the triad of AS, acquired coagulopathy (von Willebrand syndrome type 2A, or vWS-2A) and anemia due to bleeding from intestinal angiodysplasia. Clinicians should be aware of the possibility of gastrointestinal (GI) bleeding due to angiodysplasia in patients with aortic valve stenosis.

Case Report

A 75-year-old female patient with a history of hypertension, chronic obstructive pulmonary disease, coronary artery disease (CAD) and

anemia requiring a total of 30 units of packed RBC in the past four years, was diagnosed with angiodysplasia of the upper and lower GI system endoscopy, especially of the cecum and distal ileum (Fig. 1). The bleeding continued after she had undergone argon plasma coagulation. The patient presented with New York Heart Association (NYHA) class III functional capacity. Echocardiography revealed severe, calcified AS with a valvular area of 0.6 cm^2 with a mean transvalvular gradient of 52 mmHg and a left ventricular ejection fraction of 60% (Fig. 2, Video 1. See corresponding video/movie images at www.anakarder.com). Even though she had been evaluated for surgical replacement of the aortic valve, the cardiac surgeon declined the operation because of very high surgical risk (Logistic EuroSCORE 22.75%).

A stent was successfully implanted in the patient's left anterior descending artery two weeks before the TAVI. A 23 mm Edwards SAPIEN valve (Edwards Lifesciences, Irvine, California) was successfully implanted percutaneously through the right femoral artery. The procedure was performed under mild sedation. The valve placement was in appropriate position, the coronary arteries were patent at the post-procedural aortic root evaluation (Fig. 3, Video 2. See corresponding video/movie images at www.anakarder.com). Post-procedural echocardiographic assessment showed a well-functioning prosthesis with 1.9 cm^2 surface area, a mean gradient of 8 mmHg and mild paravalvular leakage. The functional capacity was class I at the 6-month follow-up. Two units of packed RBC were replaced in the first week after the procedure, after than no blood transfusion was needed. Endoscopy two months after the procedure showed a disappearance of angiodysplasia.

Discussion

Aortic stenosis may only be detected in later symptomatic stages, while intestinal angiodysplasia will not always result in anemia; routine investigations may not reveal these two underlying conditions as the cause of a patient's anemia. Therefore, the prevalence of Heyde syndrome is not clearly determined and many mild cases are likely to remain undiagnosed. Acquired vWS-2A in AS is caused by degradation of von Willebrand factor (vWF) multimers by shear stress across the diseased valve (1). Similarly, hypertrophic obstructive cardiomyopathy, ventricular septal defect and left ventricular assist devices associated with shear-induced vWF conformational changes leading to increased proteolysis of vWF (2). The deficiency of von Willebrand's factor led to a secondary reduction in plasma factor VIII level and impaired platelet adherence to subendothelial components. The development of iron deficiency anemia in an elderly patient with established AS should evoke the possibility of Heyde syndrome. Initial investigations must explore other possibilities, such as underlying gastrointestinal malignancy, celiac disease or nutritional deficiency.

Aortic valve replacement (AVR) should be recommended as a "gold standard" (3). It is important to keep in mind that lifelong anticoagulation will be required if a mechanical valve is considered for the AVR and this anticoagulation treatment will increase the possibility of GI bleeding in patients with Heyde syndrome. A biologic prosthesis is recommended for valve replacement as the first line treatment for AS patients with coexis-

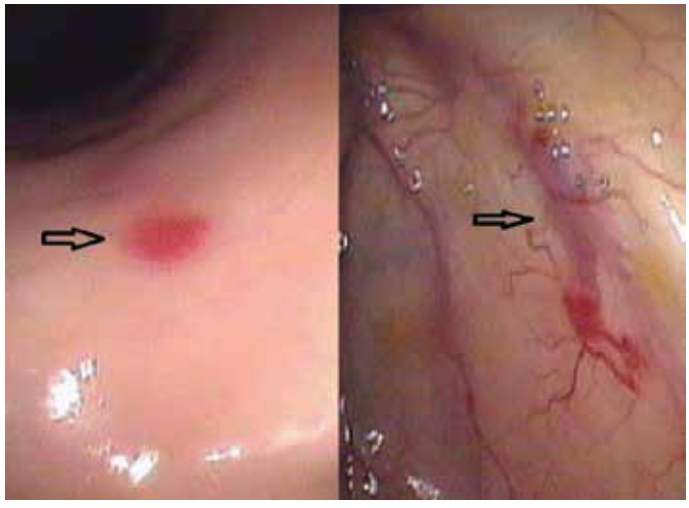

Figure 1. Location of angiodysplastic lesions in different areas of the gastrointestinal tract, marked by arrows

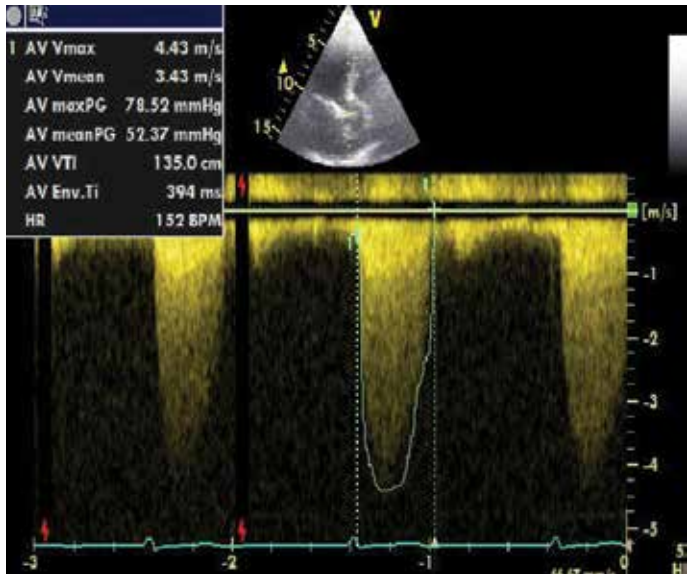

Figure 2. Pre-TAVI continuous Doppler echocardiographic image of the aortic valve

TAVI - transcatheter aortic valve implantation

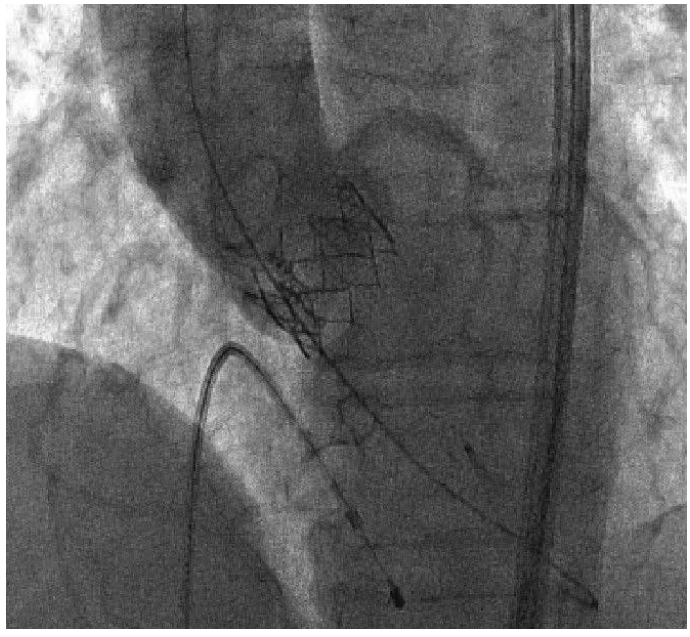

Figure 3. Image of the aortic root immediately following valve placement by TAVI

TAVI - transcatheter aortic valve implantation

tent GI bleeding (4). From the first day after surgery for valve replacement, all patients had their high molecular weight vWF multimers levels and platelet function brought to normal conditions, which proves the relationship between valve disease and hematological abnormality (5).

When valve surgery cannot be performed, for high surgical risk, authors suggest performing colectomy after identification of the bleeding site as an initial treatment option, knowing that recurrence of bleeding could occur at another site (6). Today, percutaneous prosthesis can be performed (7).

Conclusion

TAVI is an alternative therapy in patients with severe AS and high surgical risk. In high risk patients such as ours, TAVI has success as a treatment of both AS and the accompanying coagulopathy.

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Video 1. Pre-TAVI long axis transesophageal echocardiographic video of the aortic valve

TAVI - transcatheter aortic valve implantation

Video 2. Video of the aortic root immediately following valve placement by TAVI

TAVI - transcatheter aortic valve implantation

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Double covered stent closure of extracardiac Fontan multiple fenestrations

Ekstrakardiyak Fontan'da, çoklu fenestrasyonların çift kaplı stent ile kapatılması

Introduction

Fontan procedure involves placement of a fenestration to allow decompression of the systemic venous circulation. Extracardiac fenestration has proved to be effective for low venous pressure, better cardiac output. However, the fenestration results in increased right-to-left shunting, with decreased oxygen saturation and potential for embolic phenomena (1). Hence, for some patients, closure of fenestration is mandatory. Interventional closure of the extra-cardiac tunnel is safe and effective for non invasive procedures (2-6).

Case Report

A 9-year-old girl presented with left atrial isomerism, complete atrioventricular septal defect, unbalanced ventricles (with right ventricle dominance, left ventricular hypoplasia) and double outlet right ventricle, pulmonic stenosis and PDA. There was no right-sided superior vena cava and a left superior vena cava drained into the left-sided systemic venous atrium. The inferior vena cava was interrupted with hemiazygous continuation to the left superior vena cava. By 9 years of age, her medical history included the following: At the age of 2.5 left 5mm Gore-Tex modified Blalock-Taussig shunt was constructed. By 4 years of age, her saturation decreased as low as 80% and she developed clubbing of the fingers. The extracardiac lateral tunnel Fontan was constructed with bovine pericardium at the age of 5 years.

On admission, her saturation decreased to the level of 76%. Echocardiography demonstrated a patent fenestration with right to left flow and satisfactory ventricular function. At catheterization, her pulmonary-to-systemic blood flow ratio was 0.35, central venous pressure 5 mmHg, mean left atrial pressure-7 mm Hg, and left ventricular end-diastolic pressure-0 to 7 mmHg, hematocrit-45.5% and tunnel mean pressure-17 mmHg. Angiography demonstrated a right-to-left shunt through the fenestration. The fenestration was in the midline 7 mm in diameter and above this, there was another tortuous defect 3-4 mm in diameter (Fig. 1a, b). These two defects prevented occlusion test of the

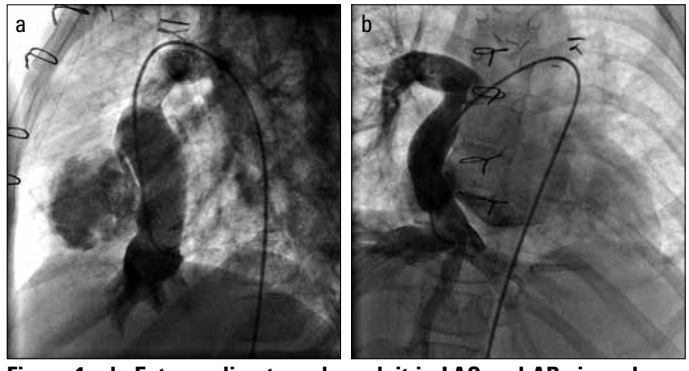

Figure 1a, b. Extracardiac tunnel conduit in LAO and AP view shows the presence of multiple fenestrations and opacification of the pulmonary atrium

LAO - left anterior oblique view, AP - anteroposterior view