

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

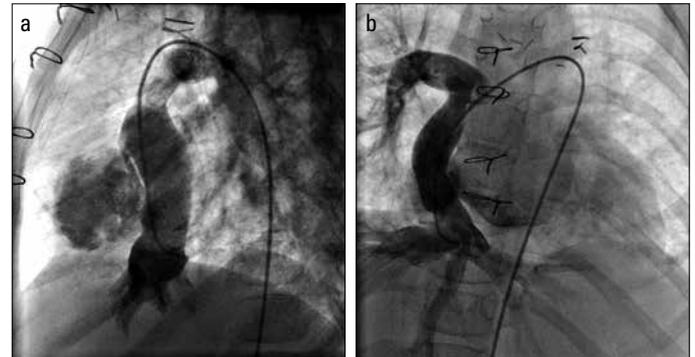


Figure 1a, b. Extracardiac tunnel conduit in LAO and AP view shows the presence of multiple fenestrations and opacification of the pulmonary atrium

LAO - left anterior oblique view, AP - anteroposterior view

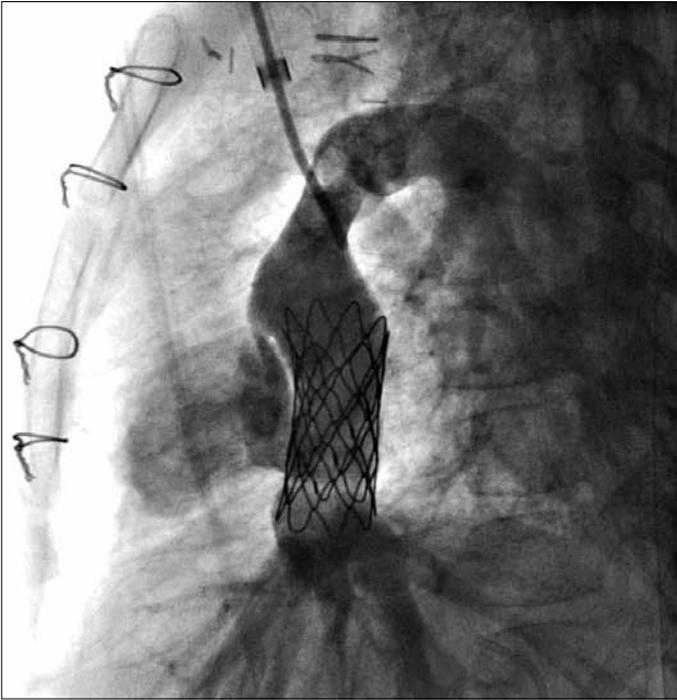


Figure 2. Cheatham-Platinum covered stent placement at central fenestration level partially abolished the right to left shunt and persistence of the second fenestration

fenestration and general anesthesia was used and orotracheal intubation with endocarditis prophylaxis 1 gr sephazolin. Intravenous heparin (100 IU/kg) was given immediately after left jugular and femoral vein cannulation. The catheter was exchanged with a standard 0.035-in., 260 cm exchange extra-stiff guide wire and 6F sheath changed with 13 French Hausdorf long sheath (Cook, Bloomington, IN, USA). The fenestration was closed with a right jugular vein access covered Cheatham Platinum stent (CP8Z39). The balloon-in-balloon delivery catheter (Numed, NY USA) was inflated at 3 atm, 22 mm diameter and 35 mm length, procedural time 75 min, and fluoroscopy time 12 min. Control angiography demonstrated complete occlusion of upper part fenestration but we also saw right-to-left shunt through the second hole of the fenestration (Fig. 2, Video 1. See corresponding video/movie images at www.anakarder.com). We decided to close second hole by same procedure. Second stent (CP8Z28) was deployed in the second hole of the tunnel. The balloon was inflated at 3 atm and 22 mm diameter. Control angiography demonstrated complete occlusion of the fenestration (Fig. 3, Video 1. See corresponding video/movie images at www.anakarder.com).

The following control echocardiography demonstrated complete closure of the fenestration.

The patient was discharged on aspirin and warfarin anticoagulant therapy. She had no clinical signs of acute femoral or jugular vein thrombosis. By the 8th month of follow up, room air saturation remained 90-93%. At control echocardiography no residual shunt through the fenestration was detected.

Discussion

Interventional extracardiac fenestration closure has been introduced as an alternative technique to surgery. Various techniques have been developed for closure extracardiac fenestration (2-6). Device

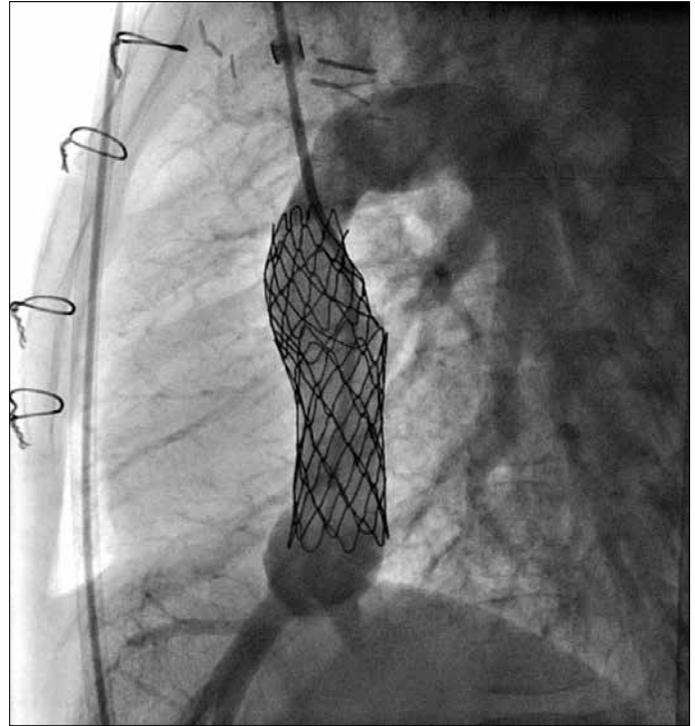


Figure 3. Second Cheatham-Platinum covered stent placement at extracardiac tunnel completely abolished the right to left shunt

choice for extracardiac fenestrations which has a large variability, is guided by several factors, including patients weight, size and location of fenestration, its geometry, vascular access and the possibility of placing a long sheath (2-6).

In our case an atrial septal defect closure device for the fenestration and a vascular plug for the 'patch leak' may be a first option. It was determined on angiography that extracardiac tunnel was dilated. There was an additional defect hole which was probably due to both suture and tunnel material failures. For this reason, we planned to use only one covered stent for safety and reliability. In our case, lower fenestration was closed with sufficient safety margins with 39 mm device but the use of the second stent was required.

Utilization the left jugular vein access in the absence of the right superior vena cava and anomalous venous return of the inferior vena cava presented the most difficulties for this patient.

Conclusion

The covered Cheatham Platinum stent is a valuable tool, and is safe. This simple and effective procedure can be used successfully for closure of extracardiac Fontan multiple fenestrations.

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Video 1. Cheatham-Platinum covered stent placement at extracardiac tunnel

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Acute aortic dissection in a 10-year-old boy with bicuspid aortic valve

Biküspit aort kapağı olan 10 yaşında erkek çocukta akut aort disseksiyonu

Introduction

Acute dissection is an unusual complication of ascending aortic aneurysm in childhood. Although progressive dilatation of the ascending aorta has been described in pediatric patients with bicuspid aortic valve (BAV), the occurrence of aortic dissection is rare (1-5).

Case Report

A 10-year-old boy presented with severe "tearing" type chest pain for 2 hours. The patient had no trauma, infection, previous cardiothoracic surgery or systemic disorder. Blood pressure and heart rate were 146/88 mmHg and 90 beats/minute, respectively. Heart sounds were diminished on auscultation. Electrocardiogram showed sinus rhythm without ischemia and low-voltage in all derivations. Chest X-ray demonstrated an enlarged mediastinum. Transthoracic echocardiography (TTE) showed dilatation of the ascending aorta and mild pericardial effusion without tamponade (Fig. 1). The diameters of the aortic annulus, sinotubular junction (STJ) and ascending aorta were 18, 28 and 44 mm, respectively. The patient was immediately transferred to intensive care unit, and transesophageal echocardiography (TEE) revealed a dissection flap above the STJ (Fig. 2, Video. See corresponding video/movie

images at www.anakarder.com). The dissection extended from the STJ to the origin of the brachiocephalic artery (BCA). The aortic arch and origins of the coronary arteries were normal. The aortic valve was bicuspid with normal function. Therefore, an emergent surgery was performed. During operation, we observed giant aneurysm of the ascending aorta (Fig. 3A). Cardiopulmonary bypass was established via the right axillary and femoral arteries, and femoral vein. After cardiac arrest and aortic incision, true and false lumens were clearly exposed (Fig. 3B). We observed that the intimal tear was above the STJ and extended up to the origin of the BCA. The aortic sinuses and coronary orifices looked normal. We explored the aortic arch during total circulatory arrest and confirmed that the dissection did not extend distally to the BCA. Supracoronary graft interposition with a Dacron graft was performed uneventfully. The patient was discharged on postoperative day 14.

At 6-month follow-up, the patient remained normotensive under medical treatment, and TTE showed normal aortic valve functions. In chromosomal analysis, there was no genetic syndrome as an underlying cause for dissection.

Discussion

The etiology of aortic dissection in children and young adults includes mostly hypertension in up to 80%, followed by BAV in 7-14% (1). The prevalence of BAV is 4.6 in 1000 live born neonates with a higher prevalence in male neonates than in female neonates (6). In a

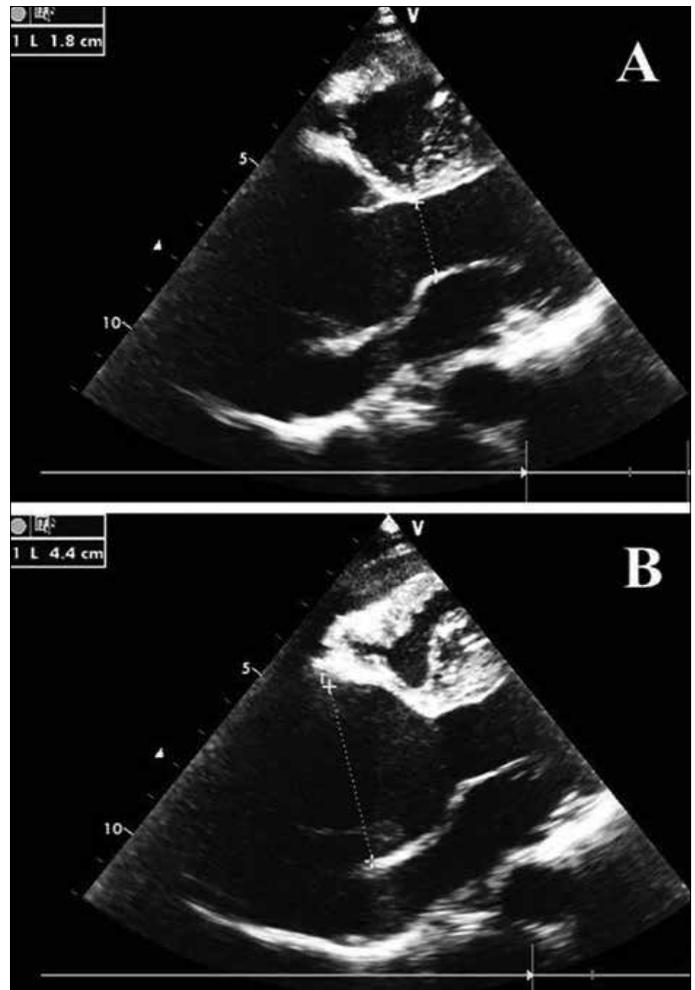


Figure 1. Transthoracic echocardiography demonstrates dilatation of the ascending aorta and pericardial effusion