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Acute thrombosis of extracardiac conduit after the Fontan operation: an emergent thrombectomy

Fontan ameliyatı sonrası ekstrakardiyak kondüitin akut trombozu: acil trombektomi

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Fontan operation is applied as palliative therapy in different type of complex congenital heart disease. Thrombosis remains a major complication after Fontan surgery, presenting as intracardiac and/or intravascular thrombosis, or other embolic phenomena. A nine-year-old boy underwent surgery to correct transposition of the great arteries, ventricular septal defect, pulmonary valve atresia by extracardiac Fontan procedure using 16 mm Dacron graft. During surgery and recovery, no adverse events were observed. On the 18th day after surgery, the patient developed sudden signs of low cardiac output, dyspnea, and right ventricular insufficiency. Angiography and echocardiography demonstrated complete occlusion of extracardiac conduit. We have performed emergent surgical thrombectomy without cardiopulmonary bypass to the patient. The postoperative period was uneventful and the patient was discharged on the 60th day.

Key words: Congenital heart disease; Fontan procedure; thromboembolism.

Fontan ameliyatı, farklı tipte kompleks doğuştan kalp hastalıklarının palyatif tedavisinde uygulanmaktadır. Tromboz, Fontan ameliyatından sonra görülen majör bir komplikasyon olup, intrakardiyak veya intravasküler tromboz ya da diğer emboli formlarında görülebilir. Ventriküler septal defekt ve pulmoner kapak atrezisi olan dokuz yaşında erkek hastaya büyük arterlerin transpozisyonunu düzeltme amacıyla 16 mm Dacron greft kullanılarak ekstrakardiyak Fontan ameliyatı uygulandı. Hastada ameliyat sırasında ve iyileşme döneminde sorun olmadı. Hastada ameliyat sonrası 18. gün ani olarak düşük kardiyak debi, dispne ve sağ ventriküler yetmezliği bulguları gelişti. Anjiyografi ve ekokardiyografi incelemelerinde ekstrakardiyak kondüitte tam tıkanma tespit edildi. Hastaya acil olarak kardiyopulmoner baypas kullanılmadan cerrahi trombektomi uygulandı. Ameliyat sonrası dönem sorunsuz geçti, hasta 60. gün taburcu edildi.

Anahtar sözcükler: Doğuştan kalp hastalığı; Fontan ameliyatı; tromboemboli.

Since its initial introduction in 1971, the Fontan procedure has undergone multiple modifications aimed at directing the systemic venous return (SVR) directly to the pulmonary artery (PA).[1] Thromboembolism is a major cause of morbidity following the modified Fontan procedure in children with univentricular heart defects. Occasionally, the hemodynamic alterations may disturb the balance between coagulation and fibrinolysis. The Fontan operation has been called paradoxical because systemic venous hypertension is imposed with concomitant PA hypotension. This hemodynamic compromise is the underlying cause of several late complications, including arrhythmia, heart failure, thromboembolism, hepatic dysfunction, and worsening cyanosis. In addition, studies of venous thromboembolic events as a primary outcome for patients who underwent the Fontan operation have noted incidence rates ranging from 3 to 16%.[2,3] Obstruction of the so-called Fontan tunnel represents a life-threatening complication and requires immediate

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intervention. Herein, we present a patient who had a surgical thrombectomy of the obstructive thrombus in the extracardiac tunnel graft.

CASE REPORT

A nine-year-old boy, who had a mean PA pressure of 10 mmHg, underwent a Fontan procedure for corrected transposition of the great arteries (C-TGA), ventricular septal defect (VSD), and pulmonary atresia. He was considered to be a high risk patient due to his mildly increased preoperative pulmonary artery pressure PAP levels; therefore, an extracardiac fenestrated Fontan operation was performed using a 16 mm Dacron conduit between the inferior vena cava (IVC) and PA. Eighteen days after the operation, the patient developed acute abdominal pain and ascites accompanied by progressive cyanosis and dyspnea. Pulse oximetry revealed an oxygen saturation (OS) of 67%, and echocardiography found possible thrombosis of the IVC and extracardiac conduit. Intravenous heparin infusion was initiated, and urgent diagnostic right-heart catheterization was scheduled. In addition, an electrocardiogram (ECG) also detected sinus tachycardia. An arterial blood gas analysis showed a baseline pH of 7.27, carbon dioxide partial pressure (pCO_2) of 38 mmHg, and oxygen partial pressure (pO_2) of 48 mmHg, and angiography displayed complete occlusion of the conduit and IVC (Figure 1). The patient's general condition deteriorated over the next two hours, and following discussion with our pediatric cardiologist, we decided that an urgent surgical thrombectomy of the extracardiac conduit was necessary. A median resternotomy was performed under general anesthesia, and heparin (5 IU/kg/hr) was given. Our patient's acceptable value for the activated clotting time (ACT) ranged between 200-250 seconds. The middle portion of the conduit was opened laterally, and the thrombus was removed using a Fogarty catheter (Edwards Lifesciences Corp., Irvine, CA, USA). During this removal, the PA was temporarily clamped around the superior anastomotic part of the graft. After sufficient blood flow through the superior and inferior parts of the graft was achieved, the thrombectomy was completed. Thereafter, the previously created fenestration was enlarged by simultaneously applying side clamps to both the graft and right atrial wall. The operation was then terminated without cardiopulmonary bypass (CPB). Finally, a high dose of intravenous heparin was given (25 IU/kg/h), and an improved OS of 80%. In the postoperative period, the patient's clinical status improved from day one, and inotropic support was terminated. The patient had an OS of 90%, and control echocardiography performed 48 hours postoperatively confirmed the absence of any clot formation and complete patency of the IVC and extracardiac conduit. Therefore, the patient was discharged from the hospital on oral coumadin (warfarin) treatment.

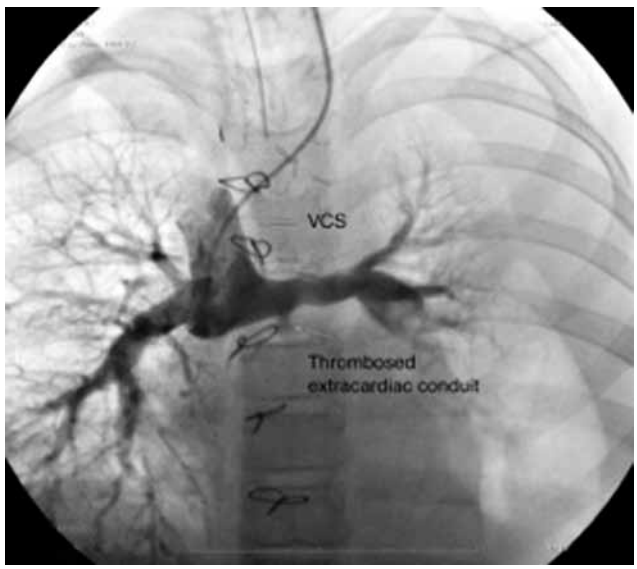

Figure 1. Angiography showing the thrombosed extracardiac conduit.

DISCUSSION

The Fontan operation has been used for a variety of congenital heart lesions and refers to a surgical intervention that bypasses the right heart. Successful surgical palliation with the Fontan procedure allows for survival into adulthood for many patients born with single-ventricle physiology, but the limited studies to date indicate that this population suffers substantial morbidity from perioperative and long-term complications, for example thromboembolic events,[3] which sometimes require intervention and ultimately a reoperation.[3] Thrombus formation can be a significant cause for morbidity and mortality after the Fontan operation. Thromboembolism after the Fontan procedure has been variously attributed to low flow states, stasis in the venous pathways, right-to-left shunts, prosthetic materials, atrial arrhythmias, and hypercoagulable states.[4] However, our patient's coagulation tests were within the normal range. The time interval between the operation and this event can vary between days and years, and the occurrence of thrombosis is always accompanied by acute clinical deterioration. The pulmonary blood flow after the Fontan operation is passive and is dependent on the transpulmonary gradient between the left atrium and the PA. Any compromise to the pulmonary blood flow, such as a decrease in the size of the pulmonary vessels,

increased pulmonary vascular resistance, or systemic ventricular failure, can lead to increased central venous pressure and eventually to sluggish right-atrial blood flow, thus producing a risk factor for venous thrombosis or thromboembolism.[5] Practices vary widely with respect to prophylactic anticoagulation strategies that have the goal of minimizing the occurrence and morbidity of thromboembolism after Fontan surgery, but there is no consensus concerning the postoperative mode and duration of anticoagulation prophylaxes. Most reports have shown scarce management and poor outcomes of thromboembolic events in patients who undergo the Fontan operation. In a study by Kaulitz et al.[3] and Cheung et al.[4] with a follow-up period ranging from one month to five years, complete resolution of thrombosis was obtained in 48% of cases and death occurred in 25%. Walker and Gatzoulis[6] reported a very poor survival rate after thromboembolic complications, with mortality rates as high as 25% in a pediatric series. In hemodynamically stable patients, thrombolysis and anticoagulant therapies can be used as initially, but these treatment strategies need at least 12-24 hours in order to be effective. In acutely deteriorating patients like ours, medical treatment may cause complications which can delay surgical treatment. Therefore, we chose the urgent surgical thrombectomy option because of the acute progressive deterioration of our patient's clinical condition. Fortunately, he dramatically improved after the surgical thrombectomy and was able to be discharged in good condition. Recently, reports have shown that optimization of the conduit is necessary on the basis of the IVC diameter. For this reason, we used a 16 mm Gore-Tex® stretch vascular graft (W.L. Gore & Associates, Medical Product Division, Flagstaff, AZ, USA) for the extra conduit.[7] Asymptomatic pulmonary emboli are frequently identified after the Fontan procedure, but we found none. In our opinion, a surgical thrombectomy should be kept in mind for patients with acute clinical deterioration after acute/subacute thrombosis of the extracardiac Fontan conduit.

Declaration of conflicting interests

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REFERENCES

1. Marcelletti C, Corno A, Giannico S, Marino B. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. J Thorac Cardiovasc Surg 1990;100:228-32.
2. Jahangiri M, Ross DB, Redington AN, Lincoln C, Shinebourne EA. Thromboembolism after the Fontan procedure and its modifications. Ann Thorac Surg 1994;58:1409-13.
3. Kaulitz R, Ziemer G, Bergmann F, Luhmer I, Kallfelz HCb Atrial thrombus after a Fontan operation: predisposing factors, treatment, and prophylaxis. Cardiol Young. 1997;7:37-43.
4. Cheung YF, Chay GW, Chiu CS, Cheng LC. Long-term anticoagulation therapy and thromboembolic complications after the Fontan procedure. Int J Cardiol 2005;102:509-13.
5. Hedrick M, Elkins RC, Knott-Craig CJ, Razook JD. Successful thrombectomy for thrombosis of the right side of the heart after the Fontan operation. Report of two cases and review of the literature. J Thorac Cardiovasc Surg 1993;105:297-301.
6. Walker HA, Gatzoulis MA. Prophylactic anticoagulation following the Fontan operation. Heart 2005;91:854-6.
7. Alexi-Meskishvili V, Ovroutski S, Ewert P, Dähnert I, Berger F, Lange PE, et al. Optimal conduit size for extracardiac Fontan operation. Eur J Cardiothorac Surg 2000;18:690-5.