Postoperative graft thrombosis in Fontan procedure

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Fontan reparir could be used as the definitive palliation in many forms of complex cyanotic congenital heart disease. But thrombosis can occur after a modified Fontan operation (right atrium-right ventricular connection with a conduit). Appropriate management of this complication includes thrombolytic therapy, thrombectomy and revision (if surgically remediable causes of the thrombosis are identified) or redo operation.

Two repair operations were performed for the treatment of thrombosis of the right side of the heart in paitents in whom we had previously performed Bjork modification (right atrium-right ventricular connection with a conduit). The thromboses occurred 6 and 9 years after the operation, respectively. In both cases, the redo Fontan operation was successfully performed using a dacron tube graft. Patients were anticoagulated after the operation. Risk of thrombosis of the right side of the heart after the Fontan repair may be minimized with the use of prophylactic anticoagulation in high-risk patients soon after the operation.

Key words: Fontan procedure, graft thrombosis, redo Fontan procedure.

Since its first introduction by Fontan and Baudet, the Fontan repair is considered the definitive palliation for patients with many forms of complex cyanotic congenital heart disease[1-3].

However, enthusiasm for and success with the one ventricle repair have been limited by our inability to prevent or adequately treat complications, such as recurrence of serous effusions, protein-losing enteropathy and thrombosis of the right side of the heart[4-11]. In this article, we address the issue of neointima formation and thrombosis in two patients, which resulted in inadequate flow through the graft between the right atrium and right ventricle, and two successful redo Fontan operations. We also review previously reported cases so as to better understand this rare but catastrophic complication.

Case Reports

Case 1

A 3.5-year-old girl was admitted to our children's hospital in 1983 for the first time with the complaints of cyanosis, fatigue and clubbing.

Her angiography revealed a large atrial septal defect, tricuspid hypoplasia, pulmonary arteriovenous fistula and hypoplastic right ventricle, and with this diagnosis it was decided the patient was a candidate for the Fontan procedure.

A modified Fontan procedure was performed on February 25, 1985 using a standard means of cardiopulmonary by-pass with a 24 mm Meadox-dacron graft (Meadox Medicals, Inc., oakland, NJ, USA).

There were no postoperative complications, and after an uneventful 15 days she was discharged with dipyridamole, digoxin, spironolactone and furosemide.

With he exception of a right bundle branch block (RBBB), her postoperative periodic controls were in normal limits till 1994, when she admitted with fatigue and edema. Complete atrioventricular (A-V) block was diagnosed and an epicardial lead and pacemaker were implanted. But her complaints did not subside, and five months later she came to the emergency unit with cyanosis, edema and ascites. Echocardiogram showed narrowing of the graft with an elevated right ventricular pressure and 10 mmHg gradient inside the baffle. The next day she was taken into surgery for a redo Fontan operation.

The redo Fontan operation was performed with right atrial-femoral cannulation and moderate hypothermia (26 °C), and a 24 mm Meadox-

dacron graft was implanted for atrio-ventricular connection (Fig. 1). The old graft material had been narrowed by neointimal proliferation and thrombus formation (Fig. 2).

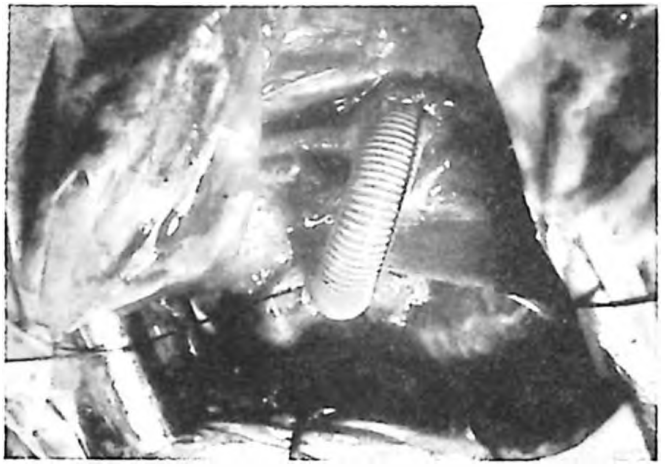

Fig. 1. Atrio-pulmonary connection with a 24 mm dacron graft at the end of the redo Fontan operation.

Fig. 2. The old graft material, which had been narrowed by the neointimal proliferation and thrombus formation.

Before hospital discharge, a control echocardiogram demonstrated patency in the graft. During her postoperative stay in hospital a protein-losing enteropathy was detected and a high protein diet was ordered (rich in protein and medium chain fatty acids). After an uneventful three weeks she was discharged. She is now free of symptoms, taking digoxin, spironolactone, and warfarin sodium, 38 months after her redo Fontan operation.

Case 2

A three-year-old boy was admitted to hospital in 1986 with cyanosis, at which time he was diagnosed with tricuspid atresia, atrial septum defect (ASD) and ventricular septum defect (VSD). With this diagnosis, a left Blalock-Taussig shunt was performed (1986).

Three years later a modified Fontan procedure was performed, and an atrio-ventricular connection was constructed using a 20 mm Meadox-dacron graft (Meadox Medicals, inc., Ocakland, NJ. USA).

After a symptom-free period of six years, he was admitted to hospital with congestive heart failure, ascites and edema. It was learned that his symptoms began four months previously and increased throughout this period.

Echocardiogram revealed narrowing of the graft, and angiography confirmed the diagnosis with an 8 mmHg gradient inside the baffle.

He was taken into surgery and a redo Fontan procedure was performed (using a 24 mm Meadox-dacron graft (Meadox Medicals, Inc., Oakland, NJ, USA) for the atriopulmonary connection. The old graft material had been narrowed by neointima and thrombus formation.

He was discharged after three weeks with warfarin sodium, digoxin he is having periodic controls and is symptom free.

Discussion

Right-sided thrombosis is a rare but fatal complication when it occurs after the Fontan procedure. In a review of the literature, we could not find many previously reported cases, and in the available clinical data, both in surgical and non-surgical interventions, the results are not satisfactory[4-13].

The data revealed that thrombosis may occur soon after the operation or may manifest itself several years after the Fontan repair (range 1 day to 15 years)[4-13].

Several factors have been implicated in predisposing a patient to thrombosis of the right side of the heart. Most of these can be grouped according to Virchow's triad of (1) stasis (2) abnormal mural surface and (3) increased viscosity or coagulability.

Systemic venous stasis may occur as a result of partial obstructions due to technical error, gradual narrowing of the anastomosis due to scarring, increased pulmonary arteriolar resistance[15,16], low cardiac output, enlarged redundant right atrium, heart block and other arrhythmias[5,15,16] or abdominal compression caused by ascites. In our patients, thrombosis

occurred after nine and six years, respectively. Although we did not observe any technical error, there was heart block and consequently ascites in one patient. Unfortunately, neither case was given anticoagulants after the first operation.

The patch or graft material used in the repair may inadvertently provide an abnormal mural surface. Central lines and shunts may provide a thrombogenic surface, which promotes thrombosis and should be removed as early as possible postoperatively. Increased blood viscosity may also contribute to formation of thrombi, as well as intravascular volume depletion and polycythemia.

Abnormalities of coagulation factors and proteins C and S and antithrombin III deficiency may develop and to formation of thrombi after the Fontan operation[7].

Diagnosis of a thrombus on the right side of the heart may be difficult. Echocardiography should be used in the routine surveillance of patients after the Fontan repair. Angiography should be performed if the diagnosis is suspected, thrombosis is unclear[4,7] or if the patient fails to make satisfactory progress after the operation. Because of the risk of embolus that may originate from the catheter tip, some groups advocate thoracal computerized tomography[18].

The best therapeutic option for acute thrombosis is unclear in the early postoperative period. If a remediable cause for the thrombosis is identified, operative intervention with thrombectomy and revision seems to be the most appropriate treatment.

Thrombolysis can be a logical alternative but it should probably be attempted only in patients who admitted early with acute thrombus formation, whose conditions are stable, who have no cantraindication for thrombolytic therapy or in whom reoperation would entail a prohibitively high risk[4].

Neither of our patients were good candidates for thrombolytic therapy as they presented with chronic symptoms.

In the early postoperative period heparinization followed by administration of warfarin may be the safest therapy currently available[5,7]. Therapeutic use fo aspirin and dipyridamole in this context has been reported only once and in that case it did not prevent recurrent thrombosis[9]. Some authors have recommended routine prophylactic anticoagulation with warfarin[5,6,13].

After the first operation, our patients were prescribed antiaggregant agents which did not preclude the thrombus formation. They were anticoagulated with warfarin sodium after the redo operations.

In our cases, we preferred performing a redo Fontan operation using a new graft material. We did not prefer thrombectomy in view of the formation of an irregular-surfaced thick neointima in both cases which might be a hazard for new thrombus formation[14].

When a patient presents with congestive heart failure, cyanosis or fatigue after asuccessful Fontan operation, one must always keep in mind the graft thrombosis and the neointima formation and should advocate an aggressive approach to diagnois: echocardiography and, if not satisfactory, cardiac catheterization. Treatment should not only emphasize removal of the thrombosis, but also should address those factors that contributed to formation of the thrombus. warfarin anticoagulation should be used prophylactically in patients who are at a high risk for thrombosis of the right side of the heart.

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