Embolic Stroke After Total Cavopulmonary Connection for Complex Congenital Heart Disease — A Case Report

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Thrombosis has been reported to occur variably from 1 day to more than 10 years after Fontan-type operation. We herein describe a case with heterotaxy syndrome with situs ambiguous, dextrocardia, single ventricle, complete atrioventricular canal defect, malposition of great arteries and bilateral superior vena cava who had received pulmonary artery banding and total cavopulmonary connection at the ages of 2 months and 4 years, respectively. However, he was lost to follow-up. Eleven years after the operation, the patient suffered from acute cerebral infarction of right middle cerebral artery territory, which was documented by brain computed tomography. Multiple venous thrombi were detected by suprasternal approach of two-dimensional transthoracic echocardiography and cardiac angiography during hospitalization. The patient finally expired in spite of aggressive treatment. We suggest that transthoracic and/or even transesophageal echocardiography should be performed as a routine surveillance for cardiac function evaluation as well as for thrombus detection in patients after cavopulmonary connection. In addition, long-term use of oral anticoagulant may be necessary, especially when patients have low cardiac output and/or high atrial pressure.

Key Words: Cavopulmonary connection • Fontan-type operation • Stroke • Cerebral infarction • Thrombosis • Anticoagulant

INTRODUCTION

The Fontan operation was firstly reported in 1971 for the management of tricuspid atresia.1 Although it has undergone a variety of modifications, the basic principle remains to divert all systemic venous blood into the pulmonary circulation without normal pumping right ventricle (RV). The most common modification is currently the total cavopulmonary connection (TCPC). It consists of a superior bi-directional cavopulmonary shunt combined with an intra-atrial or extra-cardiac lateral tunnel from the inferior vena cava (IVC) to the inferior aspect of the right pulmonary artery (PA).2 The ideal patients to receive this particular operation will be those in sinus rhythm, aged 4-15 years, and having a mean PA pressure of less than 20 mmHg and a pulmonary vascular resistance of less than 4 Wood units. In addition, the distal PA should be in adequate size, the central PA should have no major distortion, and the left ventricular ejection fraction measured by echocardiography should be at least 60%.

These operations may result in a long-term functional benefit. However, the 15-year survival rate is 55 to 80%. The causes of death are usually related to late complications.1,3 Although stroke can occur after Fontan or bi-directional Glenn’s procedure, the incidence is relatively low, occurring in approximately 2% of patients.4 It occurs in most cases in the immediate postoperative
We report herein an adolescent patient who had TCPC for his complex congenital heart disease 11 years ago and developed an abnormal shunt from intracardiac baffle and IVC through hepatic vein into the RA. Thus, it caused clinical cyanosis and finally resulted in his stroke and death.

**CASE REPORT**

The patient was a victim of heterotaxy syndrome with situs ambiguous, dextrocardia, single ventricle, complete atrioventricular canal defect, malposition of great arteries and bilateral SVC. Pulmonary artery banding procedure was performed at the age of 2 months and TCPC at the age of 4 years. He was lost to follow-up for 11 years. Acute headache and vomiting were noted for 1 day. Physical examination revealed cyanosis, clubbing fingers, single second heart sound and grade 2/6 systolic murmur over the right lower sternal edge. The neurological examination was essentially negative. Arterial blood gas analysis revealed a pH of 7.31 units, PO2 21 mmHg, PCO2 32 mmHg, and serum bicarbonate 18.5 mEq/L. The hematocrit was 46%. The serum protein C and S were decreased to 32% and 31%, respectively. Brain computed tomography (CT) revealed no significant abnormality (Figure 1A). Transthoracic echocardiography showed dextrocardia, single ventricle, complete atrioventricular canal defect, severe atrioventricular regurgitation, ejection fraction of 48%, fractional shortening of 23%, sluggish flow in SVC, and no intracardiac thrombus. During cardiac catheterization, the pulmonary arterography showed sluggish flow and the contrast medium abnormally passed through the intra-atrial baffle to dilated hepatic veins and finally draining into the RA. Several contrast-filling defects within the right PA and intra-atrial baffle were also noted (Figure 2). They were all considered as venous thrombi. All the hemodynamic and selective oxygen saturation data are illustrated in Figure 3.

After disappearance of clinical symptoms, the patient was discharged home and took aspirin for anti-thrombotic treatment. Also, he was scheduled for shunt ligation, either by surgical or percutaneous transcatheteral method. Unfortunately, 6 weeks later, sudden onset of headache, consciousness change and left hemiparesis developed. The repeated brain CT scans showed infarction of right MCA territory (Figure 1B). Being beyond the golden period as well as concern of hemorrhagic transformation of recombinant tissue plasminogen activator therapy, he received conservative treatment and warfarin for anticoagulation. In addition, intravenous mannitol and furosemide and hyperventilation method were applied to him for controlling the increased intracranial pressure. The bedside transthoracic echocardiography showed mul-

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**Figure 1.** First brain CT (A) at the level of the third ventricle with contrast medium showed essentially negative, and 6 weeks later, the repeated brain CT (B) showed the low density (arrow) at the right fronto-temporal region, compatible with acute infarction of right MCA territory.
tiple thrombi within the right SVC in spite of warfarin treatment. The patient expired from acute cerebral infarction 3 days later.

DISCUSSION

The incidence of thrombus formation was 15.3% in patients after Fontan operation. Thrombus formation could be found without clinical manifestations in about one third of patients. According to the Virchow’s triad of (1) stasis, (2) abnormal mural surface, and (3) increased viscosity or coagulability, there are several risk factors for thrombogenesis in patients after Fontan-type operation, which include the Fontan circulation itself (arrhythmias, blood flow stasis, leakage or residual right-to-left shunts); the underlying cardiac disorders (ventricular failure, atrioventricular valvular regurgitation); and other complications (endocarditis and orthopedic problems). Recent evidence suggests that increased coagulability may exist in patients after Fontan operation. Cromme-Dijkhuis et al. and Rauch et al. found that patients after Fontan operation...
had a significant decrease of protein C and S levels, and half of the patients had a higher value of coagulation factor VIII, both of which were also associated with an increase of thrombotic risk.\textsuperscript{11,12}

Thrombosis after Fontan operation is reported to exist anywhere in the circulation, such as the junction of left SVC and PA, atrial wall, interatrial septum, atrioventricular baffle, iliac vein, pulmonary vein, IVC, PA, RA, PA stump, coronary artery, cavopulmonary anastomosis region, attaching to the pacemaker leads, and the left lower lobe PA. The thrombosis occurs variably from 1 day to more than 10 years after surgery,\textsuperscript{7,10,13,14} and frequently after Fontan operation, with the first peak in the first postoperative year and second peak beyond 10 years.\textsuperscript{5}

The diagnosis of thrombus may be difficult. A thin layer of clot around the atrial wall is usually hard to recognize.\textsuperscript{14} Some thrombi have been found only at re-operation or in autopsy.\textsuperscript{7,14} Thus, a negative echocardiogram does not exclude the possibility of thromboembolism in a symptomatic patient. Several methods for detection of thrombus should be utilized. Transesophageal echocardiography has higher sensitivity than transthoracic echocardiography.\textsuperscript{4,6,13} Angiography should be performed if the diagnosis is suspected, the underlying cause of the thrombosis is unclear, or the patient fails to make satisfactory progress after operation.\textsuperscript{7,13,14} For patients after Fontan-type operation but without adequate anticoagulation therapy, periodic transesophageal echocardiography to exclude eventual thrombus was suggested.

Regarding our patient, the real cause of the first experience of headache and vomiting remains unknown. Transient ischemic accident might be a possible cause. Six weeks later, the repeated headache, conscious change and left hemiparesis were definitely resulted from the right MCA infarction. Many different therapies have been reported,\textsuperscript{5,7-13} including oral anticoagulant therapy, peripheral intravenous or catheter-directed thrombolysis with streptokinase or recombinant tissue plasminogen activator, and surgical thrombectomy. Oral anticoagulation is usually the initial treatment and widely used. However, anticoagulant therapy is not handled uniformly well in patient after Fontan-type operation.\textsuperscript{3,6,8,9} The risk of hemorrhage with long-term oral anticoagulant is regarded as acceptably low and can be reduced by modifications of therapy, frequent monitoring of blood coagulation, and careful patient selection.\textsuperscript{9,15}

In all patients after Fontan-type operation, we suggest that transthoracic and/or transesophageal echocardiography should be performed as a routine surveillance for both cardiac function evaluation and early detection of thrombogenesis. Long-term use of oral anticoagulant may also be necessary, especially when patients have low cardiac output and/or high atrial pressure.

\textbf{REFERENCE}


複雜型的先天性心臟病病患接受完全上下腔靜脈－肺動脈連接術後發生腦栓塞 — 病例報告

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Fontan 手術後可能发生血栓等併發症。其發生的時間差異相當大，可以在手術後的第一天發生，也可以在手術超過十年以後才發生。我們在此報告一位複雜型的先天性心臟病病患，他在出生二個月大時先接受肺動脈縮小手術；在四歲大時接受完全上下腔靜脈－肺動脈的連接手術。手術十一年後，病患出現頭痛症狀，但並無神經學檢查之缺陷。第一次的腦部電腦斷層檢查並無異常，心導管檢查發現有明顯的右至左分流，由下腔靜脈經肝靜脈再流入右心房。在右側肺動脈及心房內人造血管處有疑似血栓的存在。經阿斯匹林抗血栓治療及頭痛之症狀治療，病患症狀逐漸改善後返家。不幸的，病患返家六週後，突然再次出現頭痛且發生意識狀態改變及左側半身輕癱的現象。第二次腦部電腦斷層檢查發現右大腦中動脈栓塞性中風，經給予降腦壓處理及抗凝血劑藥物 (warfarin) 治療仍無效，在入院第三天後死亡。雖然 Fontan 式手術後血栓的形成並不是少見的併發症，但是，抗凝血劑使用的策略及原則，一直到現在都沒有的一致的共識。經過此案例的痛苦經驗，我們建議：(一) 任何 Fontan 式手術後的病患，都應接受長期甚至是終身的抗凝血劑治療，以期預防血栓併發症的發生，並且需要定期檢測抗凝血劑治療的效果；(二) 定期追蹤經胸前或是經食道的心臟超音波檢查，除了評估心臟功能外，也可以儘早偵測血栓的存在，以預防手術後的併發症。

關鍵詞：完全上下腔靜脈－肺動脈連接術、Fontan 式手術、單一心室、中風、血栓、抗凝血劑。