INTRODUCTION

The Fontan operation, or its iterations, is the palliative treatment of choice for most univentricular congenital heart disease.1 Thrombotic and thromboembolic phenomena are frequent and potentially serious complications after Fontan surgery; its ideal prophylaxis has not yet been fully elucidated.2-3 We present the case of a girl who underwent Fontan surgery and suffered a fatal pulmonary thromboembolism (PTE) following surgery.

CLINICAL CASE

We present the case of an 11-year-old girl with type 1b tricuspid atresia (normal large vessels, restricted interventricular communication, and pulmonary stenosis) on whom Fontan surgery was performed. When she was a neonate, a balloon atrioseptostomy followed by a right Blalock-Taussig fistula were performed. At age 4, a Glenn bidirectional shunt was placed. The patient developed increasing stress dyspnea, moderate cyanosis, and acropachy. Preoperative median arteriopulmonary pressure was slightly elevated (18 mm Hg) and there was left ventricular dysfunction (telediastolic pressure 15 mmHg; 50% ejection fraction). The Fontan procedure was completed with an 18 mm-diameter extracardiac Dacron® conduit from the inferior to superior vena cava, fenestrated with a 6-mm (Gore-Tex®) tube. After surgery thromboembolic prophylaxis with enoxaparin (Clexane®) at a dose of 1 mg/kg/12 hours was

INTRODUCCIÓN

El procedimiento de Fontan, o sus iteraciones, es el tratamiento paliativo de elección para la mayoría de las cardiopatías congénitas univentriculares.1 Los fenómenos trombóticos y thromboembólicos son frecuentes y potencialmente serios complicaciones después del procedimiento de Fontan; su ideal de profilaxis no ha sido aún completamente elucidado.2-3 Presentamos el caso de una niña que fue sometida a un procedimiento de Fontan y sufrió un tromboembolismo pulmonar (TPE) a continuación de su cirugía.

CASO CLÍNICO

Presentamos el caso de una niña de 11 años de edad con tipo 1b atresia tricúspide (vías grandes normales, comunicación interventricular restrictiva, y estenosis pulmonar) a quien se le realizó la cirugía de Fontan. Cuando era un neonato, se le realizó un atrioseptostomía con balón seguida de una fistula de Blalock-Taussig. A la edad de 4 años, se le colocó un Glenn bidireccional. La paciente desarrolló disnea de esfuerzo, moderada cianosis y acropachía. La presión media arterio-pulmonar preoperatoria fue levemente elevada (18 mm Hg) y había disfunción ventricular izquierda (presión telediastólica 15 mmHg; fracción de eyeción del 50%). El procedimiento de Fontan se completó con un conducto extracardíaco de Dacron® de 18 mm de diámetro, de inferior a superior vena cava, fenestrado con un tubo de Gore-Tex® de 6 mm. Después de la cirugía, se le administró profilaxis tromboembólica con enoxaparin (Clexane®) a una dosis de 1 mg/kg/12 horas.

Brief Reports

Pulmonary Thromboembolism after Fontan Operation

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The fatal outcome in an eleven-year-old girl, one month after an extra-cardiac Fontan operation is reported. She was diagnosed with tricuspid atresia and had a Blalock Taussig shunt and a bidirectional Glenn procedure. The Fontan operation was performed using a Dacron conduit, fenestrated with a 6 mm Goretex® tube. The first week after the operation she received low molecular weight heparin, then it was stopped and aspirin was started. One month after surgery she was admitted to the hospital because of sudden cyanosis, dyspnea, chest pain and syncope. A diagnosis of left pulmonary artery thrombosis without right to left shunt across the fenestrated tube was attempted. A local infusion of rtPA was started without improvement and she died 3 hours later.

Key words: Fontan procedure. Thrombus. Fibrinolysis.

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administered for 1 week with 5 mg/kg/day aspirin added later. Postoperatively, moderate right cardiac insufficiency with pleural hemorrhage, ascitis, and hepatomegaly were treated conservatively with a low-fat diet, diuretics, and vasodilators. One month after surgery the patient was admitted to the emergency room with sudden cyanosis, intense dyspnea, thoracic back pain, and presyncope. Helicoidal computerized axial tomography (CAT) showed a repletion defect in the left pulmonary artery that continued through the inferior lobar arteries and lingually from the thrombus (Figure 1). Cardiac catheterization was performed; on angiography complete obstruction of the left pulmonary artery was observed without a fenestration short circuit (Figure 2). We then performed mechanical lysis of the thrombus and local fibrinolysis with tPA (0.6 mg/kg/h, for 6 hours); despite which the patient died 3 hours later of cardiac shock. At autopsy complete obstruction of fenestration by a thrombus around the Gore-Tex® tube was observed, as were obstruction of the left pulmonary artery and a thrombus of the inferior vena cava and renal vein outlets.

DISCUSSION

Although the incidence of intracardiac thrombosis in the proximal veins of the heart following Fontan surgery is high (up to 33% in asymptomatic patients3,4) PTE, a serious complication with a mortality rate of approximately 50%, has rarely been described. Transesophageal echocardiography is very sensitive for the detection of intracardiac thrombus5,7 but its usefulness in diagnosing PTE has not been defined. In this patient, obstruction of the Gore-Tex® tube in
fenestration would probably have been detected, but possibly not the pulmonary artery thrombus. Helicoidal CAT rapidly confirmed the clinical diagnosis and precisely located the thrombus. There are many risk factors for thrombosis after Fontan surgery: a) anastomosis and non-biological prosthetic implants in a low pressure circulatory system, especially in this hemodynamically high-risk case, and b) The existence of a procoagulate post-operative state. In this patient, another associated factor may have been the early occlusion of fenestration and its deleterious affect on cardiac output, with major slowing of venous circulation. The point at which thromboembolic complications present varies: 50% of patients in the first 3 months and later on in the remainder (mean, 6.1 years). Effective prophylactic therapy, therefore, should be administered for more than 3 months, although recommendations vary greatly and there is no consensus regarding the type and duration of therapy.

As far as treatment of serious PTE is concerned, tPA has been successfully used after mechanical thrombosis with a modified pigtail catheter. In this patient, this option was used in the place of surgery which, although useful in some cases, was ruled out because of hemodynamic instability.

In conclusion: a) there is a high risk level of PTE following Fontan surgery; b) helicoidal CAT is useful for its immediate diagnosis, and c) prospective multicenter studies are needed to define the correct prophylaxis for this procedure.

BIBLIOGRAFÍA