Pulmonary Thromboembolism after Fontan Operation

Fernando Benito Bartolomé, Fredy Prada Martínez and Monserrat Bret Zurita[a]

Sección de Hemodinámica. Servicio de Cardiología Pediátrica. [a]Servicio de Radiología.
Hospital Universitario La Paz. Madrid.

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 made. She was carried to the cardiac catheterization laboratory where a mechanical lysis of the thrombi 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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Tromboembolismo pulmonar tras operación de Fontan

Se describe la evolución fatal de una niña de 11 años, un mes después de haber sido sometida a una operación de Fontan. Diagnosticada de atresia tricúspide se le realizó, previamente a la corrección, una fístula de Blalock-Taussig y un Glenn bidireccional. La operación de Fontan se completó con un conducto extracardíaco de Dacron, fenestrado con un tubo de Goretex® de 6 mm. Tras la cirugía, la primera semana recibió heparina de bajo peso molecular y, a continuación, aspirina. Al mes de la operación ingresó de urgencia con cianosis súbita, disnea, dolor torácico y síncope. Se diagnosticó un tromboembolismo pulmonar, en la arteria pulmonar izquierda, y se procedió a la lisis mecánica del trombo y fibrinólisis local con tPA sin éxito, falleciendo 3 h más tarde.

Palabras clave: *Procedimiento de Fontan. Trombo. Fibrinólisis.*

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.[2-5] We present the case of a girl who underwent Fontan surgery and suffered a fatal pulmonary thromboembolism (PTE) following surgery.

Correspondencia: Dr. F. Benito.
Meléndez Valdés, 22, 5.° B. 28015 Madrid. España
E-mail: fbenito@gmx.net

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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 Glen 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 18mm-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

ABREVIATURAS

TEP: tromboembolismo pulmonar
TC: tomografía axial computarizada

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 lingularly from the

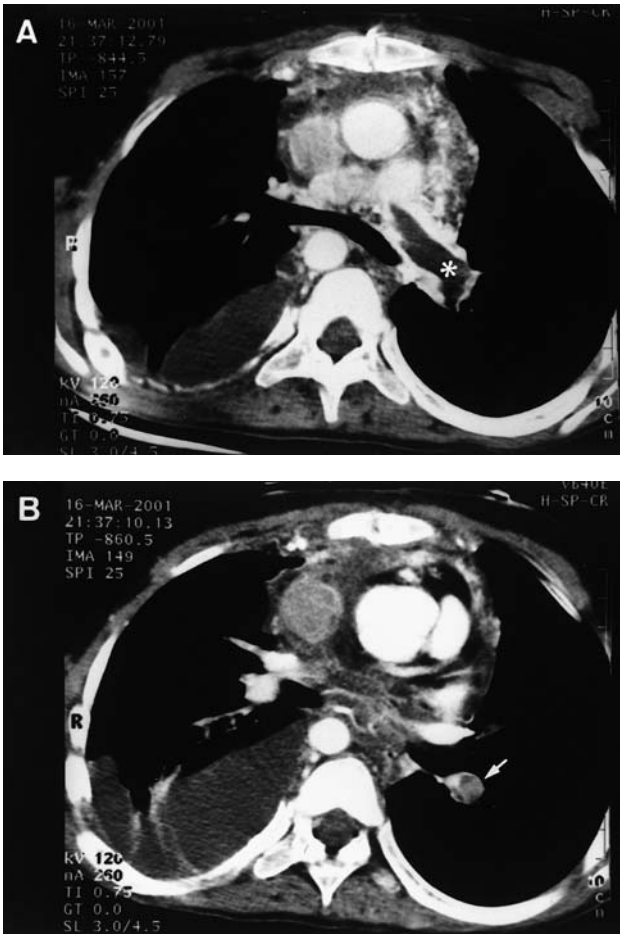

Fig. 1. Imagen de TC; cortes en la carina (A) y en un plano más caudal, en la raíz aórtica (B). En (A) se observa una imagen de defecto de repleción en el interior de la arteria pulmonar principal izquierda, alargado y adherido a la pared superior del vaso (asterisco), que se continúa por la arteria lobar inferior izquierda (flecha) (B), sugestiva de trombo. El conducto extracardíaco de Fontan está situado a la derecha de la aorta ascendente, poco contrastado, por el escaso retorno venoso de la VCI en fases precoces. Asimismo, se aprecia la presencia de un derrame pleural derecho encapsulado.

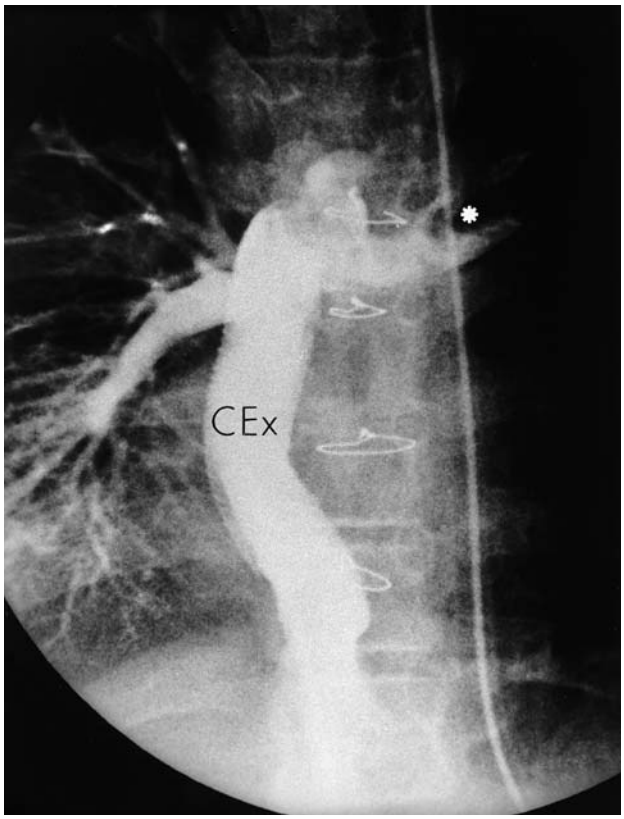

Fig. 2. Angiografía selectiva en el conducto extracardíaco de Dacron® (CEx), en proyección anteroposterior, que conecta la vena cava inferior con la superior. Se observa la obtrucción completa, por trombosis, de la arteria pulmonar izquierda (asterisco).

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 patients[3,4]) 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 thrombus[6,7] 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[3] in a low pressure circulatory system, especially in this hemodymically high- risk case, and *b)* The existence of a procoagulate post-operative state.[9] 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).[10] 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.[11]

As far as treatment of serious PTE is concerned, tPA has been successfully used after mechanical thrombosis with a modified pigtail catheter.[12] In this patient, this option was used in the place of surgery which, although useful in some cases,[5] 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.

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