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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 shunt 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 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

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administered for 1 week with 5 mg/kg/day aspirin added later. Postoperatively, moderate right cardiac insufficiency with pleural hemorrhage, ascites, 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 patients\(^{35}\)) 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\(^5,7\) 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.

BIBLIOGRAFIA