the left common carotid artery can be performed if the left subclavian artery is involved. It is also used for long segment disease. Patch ostioplasty is ideal for an isolated ostial LIMA lesion not involving the left subclavian artery [6].

High serum levels of circulating low-density lipoprotein are deposited on the aortic valve. Secondary changes such as calcification and inflammation lead to development of severe calcific aortic stenosis, sometimes extending to the mitral valve. Usually disease starts much earlier in life when the aortic root is small. Deposition of calcium along with deformed growth of the aortic root will cause a small aortic root, posing problems for aortic valve replacement. Often, the root-enlarging procedure is required to fit in an adequately sized valve [7].

The first operation was performed and reported by Dr Anil Bhan, formerly a Professor at our institute.

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FEATURE ARTICLES

Thoracic Duct Decompression for Protein-Losing Enteropathy in Failing Fontan Circulation

Marta António, MD, Andreia Gordo, MD, Conceição Pereira, MD, Fátima Pinto, MD, Isabel Fragata, MD, and José Fragata, MD, PhD

Departments of Pediatric Cardiology, Cardiothoracic Surgery, and Anesthesia, Hospital de Santa Marta, Centro Hospitalar Lisboa Central, Nova Medical School, Lisbon, Portugal

An infrequent but devastating late complication of Fontan circulation is protein-losing enteropathy (PLE), which results from unbalanced lymphatic homeostasis. Surgical decompression of the thoracic duct by redirecting its drainage to the pulmonary venous atrium has been introduced recently as a possible treatment. This report describes a single-institution experience with this innovative procedure in 2 patients with failing Fontan circulation with PLE refractory to optimized medical therapy.

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Since 1968, palliation of functional single ventricles has undergone many revisions to improve survival and decrease long-term morbidity. Fontan circulation now encompasses a spectrum of anatomical substrates, staging options, and operative techniques [1, 2]. Inherent to this circulation model is the absence of a dedicated power source to serve the pulmonary circulation and, consequently, chronic elevation of central venous pressure (CVP). This will unbalance lymphatic homeostasis, leading to fluid accumulation [2, 3]. Lymphatic insufficiency has a central role in the disease process of 1 of the most devastating complications of Fontan procedures—protein-losing enteropathy (PLE) [4, 5]. To improve lymphatic drainage, a new surgical concept based on decompression of the thoracic duct to the lower pressure levels of the systemic atrium was recently introduced [3].

We describe our experience with this innovative procedure in 2 patients with PLE refractory to optimized medical therapy for their failing Fontan circulation.

Case Reports

Patient 1

A 17-year-old boy with hypoplastic left heart syndrome underwent neonatal Norwood palliation followed by a bidirectional cavopulmonary anastomosis at 9 months of age and Fontan completion through a fenestrated extracardiac conduit at 3 years of age. The last operation was complicated by third-degree atrioventricular block treated by double-chamber epicardial pacing. Eleven years later his exercise capacity deteriorated, with severe heart failure, ascites, and hypoalbuminemia. Cardiac catheterization showed an unobstructed Fontan pathway with a mean pressure of 18 mm Hg in the main pulmonary artery; the fenestration was patent and there was no systemic outflow tract obstruction. Pulmonary vasodilator therapy achieved only slight improvement. Two years later, he underwent replacement of his pacemaker system and subsequently experienced progressive hypoalbuminemia, hypoproteinemia, and ascites despite aggressive medical therapy and multiple albumin transfusions. He was listed for heart transplantation, but because of successive positive cross-matches, the patient began a pretransplantation desensitization program with rituximab and bortezomib. Forty-eight hours after the

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Address correspondence to Dr António, Hospital de Santa Marta, Centro Hospitalar Lisboa Central, Nova Medical School, Lisbon, Portugal; email: marta.antonio@gmail.com.

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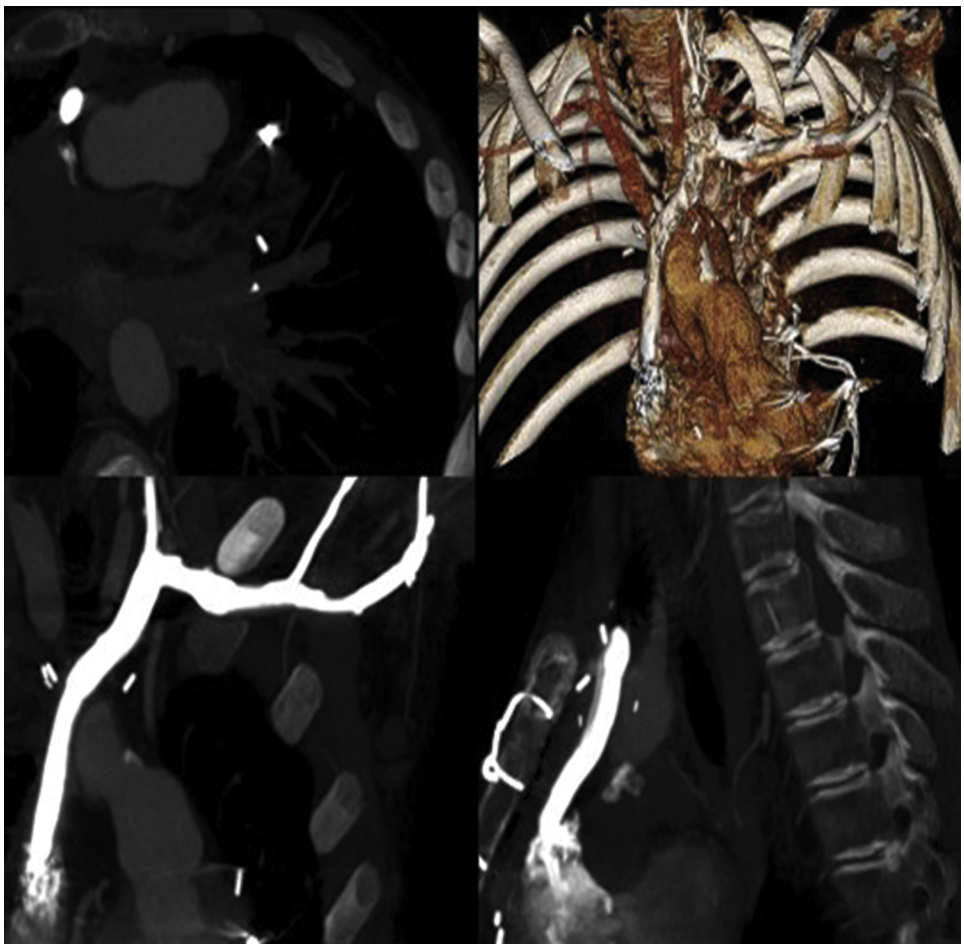

Fig 1. Computed tomographic (CT) angiography shows patent innominate vein–to-atrium conduit.

first drug cycle, he experienced vasoplegic septic shock. Despite 2 months of aggressive medical therapy and weekly paracentesis, his clinical status did not improve, and he was unable to be weaned from intravenous inotropic agents. Being at a therapeutic impasse, surgical decompression of the thoracic duct was performed by diverting the innominate vein to the atrium through an 8-mm Gore-Tex (W. L. Gore Associates, Flagstaff, AZ) graft. Ascites continued for another 5 weeks but then progressively resolved with significant clinical improvement. He was discharged 7 weeks after the operation but later presented with facial edema. Computed tomographic angiography (CT angiography) revealed a patent innominate-atrial anastomosis (Fig 1). By 10 months after the surgical procedure, he was weaned from corticosteroids, with complete resolution of facial edema, a normal serum albumin level, arterial saturation of 88%, and only mild ascites (Table 1). Clinically, the patient remains dramatically improved and his condition is stable.

Patient 2

A 15-year-old girl with a 9-year history of PLE underwent the same operation while awaiting transplantation for her failing Fontan circulation. Born with a double-inlet left ventricle, transposition of the great arteries, and coarctation of the aorta, she underwent coarctation repair and pulmonary artery banding in the neonatal period, followed by a Damus-Kaye-Stansel anastomosis and bidirectional cavopulmonary anastomosis 8 months later. At 3 years of age, the Fontan circulation was completed by a fenestrated extracardiac conduit. This was complicated by third-degree atrioventricular block, which was treated by double-chamber epicardial pacing. Twelve years later she experienced severe ascites, facial edema, and persistent hypoalbuminemia; Fontan pathway mean pressure was 15 mm Hg at cardiac catheterization, but there was sluggish

Table 1. Comparison of Clinical Data Before and After Operation

Clinical Data	Patient 1 Before Operation/ After Operation		Patient 2 Before Operation/ After Operation	
Serum albumin	28 g/L	>35 g/L	18 g/L	>35 g/L
Ascites	Severe	Mild	Severe	Mild
Facial edema	. . .	. . .	Moderate	Mild
Arterial saturation	94%	88%	99%	94%

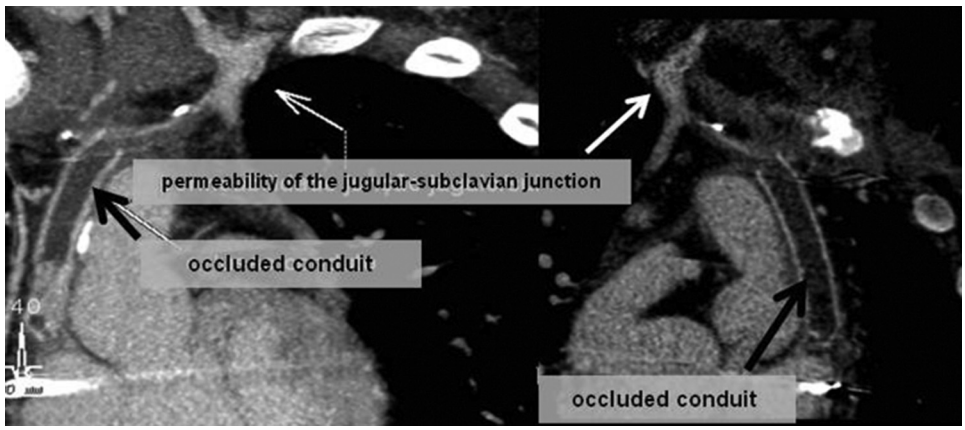

Fig 2. Computed tomographic (CT) angiography shows completely occluded innominate vein–to-atrium conduit.

flow in the inferior vena cava, and the fenestration was not patent. She had no systemic outflow tract obstruction. Aggressive medical therapy with 2 pulmonary vasodilators, low-molecular-weight heparin, selective oral corticosteroids, multiple albumin transfusions, and intravenous diuretics did not improve her clinical status significantly despite a slight improvement in albumin levels. Thoracic duct diversion was complicated by right pleural effusion, but she was discharged 4 weeks later. Control CT angiography 4 months later revealed complete occlusion of the innominate-atrial anastomosis (Fig 2). At last follow-up 6 months after the surgical procedure, the patient was still receiving oral corticosteroids, arterial saturation ranged from 94% to 95%, and both ascites and facial edema had improved (Table 1). She remains hemodynamically stable with normal serum albumin and protein levels and no recurrence of pleural effusions.

Comment

Management of late Fontan complications, particularly PLE, remains challenging. This secondary disorder is characterized by profound loss of serum proteins into the gastrointestinal lumen caused by enteric mucosal injury, systemic inflammation, and venous or lymphatic obstruction [4].

The relative infrequency of PLE, combined with our rudimentary knowledge of the interplay between the cardiovascular and lymphatic systems, has hindered understanding of the pathophysiologic processes underlying this disease, as well as the development of effective therapy. Transplantation is often the only reasonable option [5, 6]. Historically, survival after diagnosis has been 50% at 5 years [4, 7]. Nevertheless, a recent study reported 88% and 72% survival rates at 5 and 10 years, respectively, after diagnosis. This markedly improved survival may reflect earlier diagnosis from routine screening [8].

Supportive evidence suggests that the lymphatic system operates at or near its physiologic limit in patients with Fontan circulation [5, 7]. Inherent to this circulation are passive pulmonary blood flow and elevated CVP, which is transmitted backward to the hepatic and intestinal venous system. This causes increased lymph production, for which the lymphatic pump cannot compensate. In addition, chronic elevation of CVP retards lymphatic return to the central venous system, thus unbalancing lymphatic homeostasis [2–5]. The procedure of thoracic duct decompression was introduced to unburden the thoracic duct and increase the transport capacity of the lymphatic system. This should improve not only the systemic venous congestion but also preload of the single ventricle, and therefore cardiac output at the expense of mild desaturation consequent to a right-to-left

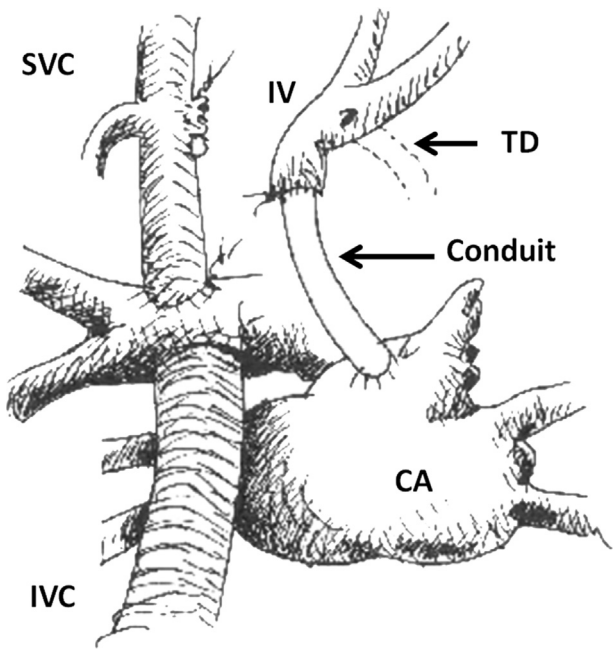

Fig 3. Interposition of prosthetic conduit (8-mm Gore-Tex graft) between the innominate vein and the atrium. (CA = common atrium; IV = innominate vein; IVC = inferior vena cava; SVC = superior vena cava; TD = thoracic duct.)

shunt [3]. Both the concept and the technique of diverting the innominate vein into the lower-pressure atrium are simple. Ideally, the largest possible direct anastomosis is preferred. However, taking into consideration that our patients had several redo operations, this fact creates challenging scenarios making it safer to interpose a prosthetic conduit because of length and technical difficulties (Fig 3). Although this not be the ideal, it may work equally well.

Our experience with this procedure in 2 patients with failing Fontan circulation who had been listed for heart transplantation suggests that 3 to 4 weeks are necessary to achieve normal serum albumin levels and improved clinical status. Arterial desaturation was acceptable after the operation. Despite receiving therapeutic low-molecular-weight heparin and platelet aggregate inhibitors, the innominate vein–to-atrium conduit occluded in 1 patient. Nevertheless, her serum albumin levels and clinical status remained improved, suggesting that some degree of lymphatic decompression, possibly through development of venous collaterals, may still have taken place. The conduit is patent in the other patient 10 months after the operation; he is receiving anticoagulation therapy with warfarin.

Although we have not yet performed transplantation in either of these patients (this being 1 of the merits of this procedure), we do not anticipate additional technical difficulties. We will either leave the connection closed, hoping that the acquired collateralization will make pressures in the system low (which can be easily measured on completion of the transplantation procedure), or conduit reconstruction to the superior vena cava system will be considered. Having said this, the status created by a full normal heart and circulation will be sufficient to deal with an eventual increase in the lymphatic drainage; however, this will have to be confirmed.

In conclusion, thoracic duct decompression for PLE is based on physiologic principles. It is technically a simple procedure despite being a reoperation. Global experience is still limited, but early results in a small number of patients have been encouraging. More experience is needed to confirm these results, elucidate optimal methods of postoperative anticoagulation, and improve understanding of the underlying pathophysiologic characteristics. Modifications of surgical technique may also offer an opportunity for improved patency of the lymphatic-to-venous connection.

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Cardiopulmonary Bypass Strategy for a Cyanotic Child With Hemoglobin SC Disease

Kelly A. Machovec, MD, MPH,
Robert D. B. Jaquiss, MD, David D. Kaemmer, CPP, FPP,
Warwick A. Ames, MBBS, FRCA,
Hercilia M. Homi, MD, PhD,
Richard J. Walczak, Jr, CPP, FPP, Andrew J. Lodge, MD,
and Edmund H. Jooste, MBChB

Department of Anesthesiology, Division of Pediatric Anesthesia; Department of Surgery, Division of Cardiovascular and Thoracic Surgery; Department of Perfusion Services, Duke University Hospital, Durham, North Carolina

Hemoglobin SC (HbSC) disease is a hemoglobinopathy that may produce sickling under conditions of hypoxemia, dehydration, and acidosis. We present a case of HbSC disease and tricuspid atresia, type IB. We describe management by cardiopulmonary bypass CPB using exchange transfusion at initiation of bypass and fractionation of collected blood, allowing platelet and plasma apheresis, as an option for patients unable to undergo this procedure off pump.

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Hemoglobin SC (HbSC) disease is a hemoglobinopathy defined by abnormal HbS and HbC in a 50:50 ratio in the erythrocyte [1]. HbC is produced by a genetic mutation causing upregulation of the potassium chloride cotransporter, increasing potassium efflux from erythrocytes and producing cell dehydration [1]. The combination of dehydration and HbS polymerization is responsible for the clinical manifestations of HbSC disease, including pain crises, acute chest syndrome, splenic

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Address correspondence to Dr Machovec, Duke University Medical Center, Box 3094, Durham, NC 27710; email: kelly.machovec@duke.edu.

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