pulmonary veins. The mean Glenn pressure was 15 mm Hg, and transesophageal echocardiography showed unobstructed pulmonary veins. There was extensive cellular ingrowth into the stent at the vertical vein end, but the pathway for pulmonary venous flow remained open. He had initially poor ventricular function and severe atrioventricular valvular regurgitation and cyanosis (oxygen saturation in the high 60s to low 70s). A positive bubble study led to cardiac catheterization 7 days postoperatively. The Glenn pressure was 12 mm Hg with a transpulmonary gradient of 5 mm Hg and right ventricular end diastolic pressure of 8 mm Hg. The Glenn circuit was unobstructed but severe stenosis was present in the left upper pulmonary vein, although with good drainage from the left middle and lower pulmonary veins and right veins. A venous collateral was coil occluded with subsequent increase in saturation from the 50s, while intubated, to greater than 70%, when extubated the next day. He was extubated 9 days after the Glenn procedure and discharged 23 days after surgery. His oxygen saturation at discharge was 75% to 80% in room air. At 9 months of age, the oxygen saturation was 88%, and echocardiography demonstrated good ventricular function, moderate atrioventricular valvular regurgitation, unobstructed right upper and

lower and left lower pulmonary veins. The mean gradient across the left upper pulmonary vein was 5 mm Hg.

We believe delaying repair of the TAPVD until the Glenn procedure permitted survival of this patient, albeit with the left upper pulmonary vein stenosis. Although an uncovered stent was used, we believe a seal around the stent was made by the cuff of tissue created around the stent when it was inflated combined with the adhesions around the heart after the previous shunt surgery. We remain vigilant about his pulmonary veins, although this has been a good early outcome.

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A new method to fenestrate the Fontan circulation

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Fenestration of the Fontan circulation helps maintain cardiac output while reducing venous and lymphatic pressure, but at the expense of arterial saturation. Although some teams perform it routinely to reduce the amount and duration of cavity transudate, the majority resort to it selectively.¹

The conventional fenestration involves a 4-mm connection between the Fontan tunnel (intracardiac or extracardiac) and the right atrium created using a kissing technique. Some

fenestrations close spontaneously, but many are obturated secondarily by a diabolo-shaped device inserted percutaneously.² This report describes a new technique of fenestration using an innominate vein to right atrial appendage shunt that can be performed post-cardiopulmonary bypass (CPB) under reliable hemodynamic conditions, offering more possibilities of late percutaneous flow regulation.

CLINICAL SUMMARY

All 27 patients receiving this type of fenestration since its first realization in January 2005 were included (2005–2010). Informed consent and institutional review board approval were obtained. At the beginning of this experience, the classic kissing fenestration was also occasionally used until we had the impression that the new fenestration technique was more appealing.

The median age was 30 (22–191) months, and median weight was 13 (10–50) kg. The driving ventricle was a single left ventricle in 14 patients, right ventricle in 11 patients, and biventricle in 2 patients.

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication June 12, 2011; revisions received Sept 5, 2011; accepted for publication Dec 8, 2011; available ahead of print March 15, 2012.

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J Thorac Cardiovasc Surg 2012;144:273-5
0022-5223/\$36.00

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doi:10.1016/j.jtcvs.2011.12.057

FIGURE 1. Fenestration of the Fontan circulation, with a shunt between the innominate vein and the right atrial appendage.

After the normothermic CPB used for the extracardiac staged Fontan is weaned, the patient's hemodynamic situation is assessed. If the transpulmonary gradient is less than 8 mm Hg, no fenestration is considered. If the gradient is more than 12 mm Hg, a fenestration is performed. Between these 2 values, the decision is made individually on many secondary criteria, such as the anatomy of pulmonary arteries, the function of the atrioventricular valves, and the function of the laboring ventricle.

The fenestration is performed after weaning from CPB, using a 6-mm polytetrafluoroethylene graft during the time of anticoagulation reversal. The graft is inserted end to side on the innominate vein and then onto the right atrial appendage (Figure 1) using tangential clamps. At times, the innominate vein is also crossclamped with 2 thin vascular clamps. In 2 patients in whom the right atrial appendage could not be freed sufficiently for tangential clamping, a short second run on bypass was initiated for the performance of this anastomosis. On completion of the atrial anastomosis, the graft is deaired and opened. Venous pressure usually decreases by 2 to 3 mm Hg. The arterial saturation also decreases but varies over a wider range from patient to patient. If the arterial saturation decreases less than 85%, the diameter of the shunt is reduced by a series of small vascular clips. The shunted flow in the right atrium can be checked by transesophageal echocardiography.

In 2 patients, in whom the initial gradient was 10 mm Hg and no fenestration had been primarily performed, the fenestration was performed on the first and second postoperative days.

Follow-up was complete with a median time of 41 (5–58) months. An angiogram was performed on follow-up to evaluate or adjust the shunt when clinically indicated (low arterial saturation or high venous pressures). As such, 10 patients received an angiogram for various reasons, 6 of whom required shunt assessment. The patients were discharged with oral anticoagulation, warfarin (Coumadin, Bristol-Myers Squibb Co, Princeton, NJ) for 6 months, followed by a lifelong antiplatelet therapy.

RESULTS

The transpulmonary gradient decreased from a median of 11 (10–12) to 9 (8–9) mm Hg. The arterial saturation remained greater than 85% in all but 3 patients. In these patients, the shunt size was reduced with clips. There was no case of postoperative bleeding.

One patient died postoperatively after a difficult course with multiple infections and eventually multiple organ failure. Although the fenestration remained open, cardiac output remained borderline until intractable infections occurred. The take-down of the Fontan circulation did not restore better hemodynamics. The other patients had a postoperative course typical for this passive circulation. The pleural drains remained in position for a median of 4 (2–8) days, and the intensive care unit stay was 5 (3–12) days.

The fenestration, followed echocardiographically in all patients and angiographically in 10 patients, did not close spontaneously. The fenestration was closed percutaneously in 5 patients and adjusted in 5 patients. The closure was obtained with an Amplatzer vascular plug (AGA Medical Corp, Plymouth, Minn) after a mean duration of 17.8 months (range, 7–48 months). In 4 patients in whom it was thought that the fenestration was still needed but shunted too large an amount of blood (increase of pulmonary artery pressure >15 mm Hg under balloon test occlusion during catheterization), the shunt was reduced using an Amplatzer Vascular Plug (Figure 2), which was partially reopened by inserting a 3.5-mm stent, after a mean duration of 4.75 months (range, 1–12 months). One patient with chylous effusion and Fontan pressure greater than 16 mm Hg had a stenosis at the shunt to atrial anastomosis (probably due to trabeculae). Implantation of a stent across the anastomosis led to a decrease in venous pressure and resolution of the chylothorax. These calibrated shunts were still open at follow-up. Except for 1 patient with multiple aortopulmonary fistulae, the arterial saturation was greater than 90% in all patients.

DISCUSSION

With most of the conventional techniques of fenestration described,³ the amount of shunting is difficult to predict and

FIGURE 2. Selective angiography in the innominate vein 4 weeks after Fontan completion and implantation of a 6-mm shunt in a 2.5-year-old child. A, Large shunt with excessive contrast in the right atrium. B, Percutaneous shunt reduction by implantation of an Amplatzer vascular plug (AGA Medical Corp, Plymouth, Minn) (10 mm) and fenestration with a 3.5-mm coronary stent.

regulate postoperatively. In comparison, our technique presents significant advantages. It is performed after weaning of CPB, when the hemodynamic situation can be reliably assessed and dictates it. It can even be performed later if the evolution of the hemodynamics remains a concern. In

addition, this fenestration allows a more precise calibration of the shunt in the operating room or later.⁴

This art of fenestration presents few other theoretic advantages. It shunts blood coming from the upper body and allows the complete inferior vena caval return, enriched by the supposed “hepatic factor,” to flow through the lungs with the presumed preventive effect against the late development of pulmonary arteriovenous fistulae.⁵ This fenestration potentially reduces the risk of paradoxical embolic events because these emboli are more prone to arise from the pelvis and lower extremities. Finally, this fenestration is easy to close interventionally and, contrary to many other methods, does not leave protruding foreign materials within the Fontan vessels or atrium, thus reducing the risk of flow disturbances and dangerous thromboembolism.

The potential disadvantages of this fenestration include its additional cost, the possible creation of tension on the innominate vein with the patient’s growth, and the variation of the amount of shunted blood with the orthostatic position of the patient. To avoid a subsequent distortion of the innominate vein with growth, we allow some redundancy to the graft (furthermore, this problem has not happened so frequently with the modified Blalock–Taussig shunts, a situation similar to that of our technique). Because this fenestration diverts blood from the upper body, especially the left upper extremity to the systemic circulation, the use of intravenous lines on the left upper extremities should be restricted and bubble filters inserted.

CONCLUSIONS

The described innominate vein to right atrial appendage shunt to electively decongest the Fontan circulation can be performed even after termination of cardiopulmonary bypass, under reliable hemodynamic conditions. It offers possibilities of late percutaneous flow regulation and closure. It has hence become our technique of choice to fenestrate the Fontan circulation in selected cases.

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