Figure 1. Intraoperative photograph demonstrating position of Starfish retractor on cardiac apex before decortication of visceral pericardium. Previously resected sections of parietal pericardium shown are also shown.

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Pulmonary venous pathway obstruction from recurrent restriction at atrial septum late after Fontan procedure

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Long-term outcome of patients after the Fontan procedure is currently excellent.¹ Despite initial reports with high incidence of postoperative obstruction of pulmonary venous pathway,² this complication has become rare.^{3,4} When it does occur, it can cause severe dete-

rioration of the cavopulmonary circulation,^{2,3} and reoperation is usually necessary. We report our experience with 3 patients presenting with this lesion late after cavopulmonary connection, with the aim of better understanding the potential causes of this complication. We also discuss methods of prevention and treatment.

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Patients and Methods

Between January 1984 and November 2002, a total of 1092 patients with single-ventricle physiology underwent Fontan procedures at Children's Hospital Boston. Among these patients, 268 were admitted for reoperation, including 3 patients with the diagnosis of isolated pulmonary venous pathway obstruction 8, 23, and 42 months after the Fontan procedure. Patients 1 and 3 had a diagnosis of hypoplastic left heart syndrome, whereas patient 2 had diagnosis of heterotaxy syndrome with situs inversus, double-outlet right ventricle, and unbalanced atrioventricular canal with pulmonary stenosis. None of the patients had obstructive pulmo-

Figure 1A. Intraoperative transesophageal echocardiogram and color Doppler (short-axis view). Preoperative echocardiogram shows turbulence at pulmonary venous outflow level, next to Cardioseal device. CA, Common atrium; PVO, pulmonary venous outflow.

Figure 1B. Intraoperative transesophageal echocardiogram and color Doppler (short-axis view). Postoperative echocardiogram demonstrates linear flow with absence of turbulence, as evidence of relieved obstruction. CA, Common atrium; PVO, pulmonary venous outflow.

TABLE 1. Preoperative data

Case	Diagnosis	Age at Fontan (mo)	Time to reoperation (mo)	Type of Fontan	Additional procedure	Symptoms	Echocardiographic LA-RA gradient (mm Hg)	Catheterization pressures (mm Hg)			
								Fontan baffle	LA	RA	End diastolic
1	HLHS (MS/AS)	25	8	LT, 4-mm fenestration	—	PLE	12	18-20	16	10	11
2	DORV/CAVC in HTX	43	23*	LT, 4-mm fenestration	TV plasty	Fatigue, cyanosis	8*	24-26	20	—	12
3	HLHS (MS/AS)	38	42	LT, 4-mm fenestration	—	PLE	7	16-23	13	8	8

LA, Anatomic left atrium; RA, anatomic right atrium; HLHS (MS/AS), hypoplastic left heart syndrome (mitral stenosis/aortic stenosis variant); LT, lateral tunnel; PLE, protein-losing enteropathy; DORV/CAVC, double-outlet right ventricle/common atrioventricular canal; HTX, heterotaxy; TV, tricuspid valve.

*Four months after closure of baffle fenestration with Cardioseal device.

nary venous drainage at initial diagnosis. All had undergone lateral tunnel cavopulmonary connection with a 4-mm fenestration. Patient 2 underwent closure of the fenestration with a percutaneous device (CardioSEAL, NMT Medical, Boston, Mass) 4 months after the Fontan operation (Table 1).

Results

Symptoms at readmission included peripheral edema and protein-losing enteropathy in 2 cases and easy fatigability and sporadic cyanotic episodes in the other. Two-dimensional echocardiography demonstrated restrictive interatrial communication in all cases (mean transatrial pressure gradient 8.6 mm Hg). Patient 2 had a discrete left atrial membrane, resulting in additional obstruction to pulmonary venous pathway (Figure 1). Hemodynamic studies showed transatrial pressure gradients of 6 and 5 mm Hg in patients 1 and 3, whereas in patient 2 a pressure gradient of 10 mm Hg was measured across the left atrial membrane (Table 1).

All patients underwent elective reoperation. After resternotomy and with cardiopulmonary bypass, the coronary sinus was unroofed posteriorly to the left atrium, and residual interatrial septum (patients 1 and 3) or fibrotic tissue at the interatrial communication level (patient 2) was resected. The postoperative course was uneventful for patients 2 and 3, whereas patient 1 had persistent pleural effusions. Patients were discharged to home on postoperative days 26, 5, and 4, respectively.

At short-term follow-up (17, 2, and 13 months, respectively), all patients have improved level of activity. There has been complete resolution of protein-losing enteropathy in patients 1 and 3. Two-dimensional echocardiography in patients 1 and 3 has shown an unobstructed pulmonary venous pathway.

Discussion

Long-term results after lateral tunnel-type Fontan connection are reported to be excellent, with an overall survival of 91% and

freedom from failure of 92% at 10 years.¹ Obstructed pulmonary venous pathway was described as an early complication of the lateral tunnel cavopulmonary connection as a result of improper baffle creation.² Technical improvements have reduced this complication, and in our experience it is rare. Previous reports^{3,4} underline the relative greater frequency of this complication in heterotaxy syndrome when associated with anomalous pulmonary and systemic venous connections, which can make surgical repair complicated with the need to create a tortuous and potentially obstructive pathway. Despite this concern, a recent report from our institution⁵ has shown excellent outcome with cavopulmonary connection in patients with heterotaxy syndrome and anomalous pulmonary venous return. This is likely due to improved surgical technique and use of extracardiac conduit in cases with complex atrial and venous anatomy (18%).

None of the 3 patients had evidence of obstruction at the pulmonary venous pathway early after the Fontan procedure, as determined by 2-dimensional echocardiography. Pulmonary venous obstruction developed slowly through a period of several months to years. In patient 2, pulmonary venous obstruction developed 4 months after device closure of the fenestration, suggesting that the additional scarring from device implantation contributed to the obstruction. The obstruction was due to fibrotic tissue at either the interatrial septum or in the interatrial course of the pulmonary veins (patient 2).

To prevent this complication, and to ensure a wide open pulmonary venous outflow, particularly in patients with mitral atresia or stenosis, we currently enlarge the communication between the pulmonary veins and the right atrium by unroofing the coronary sinus posteriorly into the left atrium, in addition to resection of the interatrial septum.

In conclusion, late onset obstruction of pulmonary venous pathway after lateral tunnel cavopulmonary connection is a rare but serious complication that may be prevented by unroofing the coronary sinus at the time of Fontan procedure.

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Complex double-outlet right ventricle repair in a neonate with complete tracheal agenesis

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Tracheal agenesis is a rare malformation with fatal consequences in very early life. We report the case of a baby girl with tracheal agenesis who during early infancy underwent complete repair of her associated {S,D,D} double-outlet right ventricle with supracardiac total

anomalous pulmonary venous connection and is still alive at the age of 10 months.

Dr Oppido

Clinical Summary

The patient was born spontaneously at a gestational age of 35 weeks. Birth weight was 2 kg, and Apgar score at 1 minute was 2, increasing to 6 after 2 minutes of mask ventilation. Critical clinical conditions required intubation, but the tube failed to pass the larynx under direct laryngoscopy. A blind end of the larynx was visualized with the aid of a flexible endoscope, whereas the tracheal bifurcation could be visualized from the distal esophagus through a tracheoesophageal fistula. A Portex 3 tube (Portex, Inc, Keene, NH) was positioned through the esophagus into the fistula, achieving a satisfactory bilateral ventilation.

Helical TC scan showed complete agenesis of the trachea (Floyd type II) with a distal tracheoesophageal fistula (Figure 1). Echocardiography showed {S,D,D} double-outlet right ventricle (with subaortic ventricular septal defect), total anomalous pulmo-

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