

Protein-Losing Enteropathy Seven Years after Total Right Ventricular Exclusion Procedure for Arrhythmogenic Right Ventricular Dysplasia

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A 27-year-old man who was diagnosed with arrhythmogenic right ventricular dysplasia (ARVD) underwent the total right ventricular (RV) exclusion procedure: the RV free wall was resected and an extracardiac total cavopulmonary connection (TCPC) was created using a 24-mm-diameter polytetrafluoroethylene (PTFE) graft. After an uneventful period of 7 years, he began to develop protein-losing enteropathy (PLE), which was resistant to medical therapy, moderate mitral regurgitation (MR) and right femoral arteriovenous fistula due to heart catheterization. Therefore, fenestration of TCPC, mitral annuloplasty and ligation of arteriovenous fistula were performed. After the operation, symptoms and levels of total protein and albumin were immediately improved and normalized. (Ann Thorac Cardiovasc Surg 2006; 12: 207–9)

Key words: total cavopulmonary connection, complication, protein-losing enteropathy, Fontan

Introduction

Protein-losing enteropathy (PLE) is a life-threatening complication after the Fontan procedure that often resists treatment.^{1,2)} We describe a case of PLE after the procedure of right ventricular (RV) exclusion, which was first reported in 2002.^{3,4)}

Case Report

A 27-year-old man first experienced syncope and ventricular tachycardia (VT) in 1995. He was given a diagnosis of arrhythmogenic RV dysplasia (ARVD) and the earliest recorded activity of VT was located in the RV free wall. Cardiorespiratory failure developed as a result of recurrent VT, and the patient subsequently required mechanical ventilation, intra-aortic balloon pumping and

extracorporeal membrane oxygenation.

Therefore, he underwent the total RV exclusion procedure in 1996, as reported previously.^{3,4)} The entire RV free wall, including the tricuspid valve apparatus, was excised under cardiopulmonary bypass (CPB). An atrial septal defect was created, and the coronary sinus was roofed with the trimmed right atrium (RA) flap to reroute blood flow into the left atrium (LA). An extracardiac total cavopulmonary connection (TCPC) was performed using a 24-mm-diameter polytetrafluoroethylene (PTFE) graft. The defect of the RV free wall was covered with a small 1-mm-thick PTFE patch and a permanent pacemaker (DDD) was implanted.

The post-operative course was uneventful and he had had a good quality of life till 2003 (7 years after the surgery), when he developed diarrhea, leg edema and decreased levels of total protein and albumin. Tc-99m labeled human serum albumin scintigraphy showed that protein was lost from the small intestine. He was diagnosed as PLE and medical treatment was tried using diuretics, heparin and steroids without success until 2004. Echocardiography revealed severe mitral regurgitation (MR) with slight prolapse of the anterior mitral leaflet (Fig. 1) and moderate left ventricular (LV) dysfunction (Table 1). Cardiac catheter examination showed reduced

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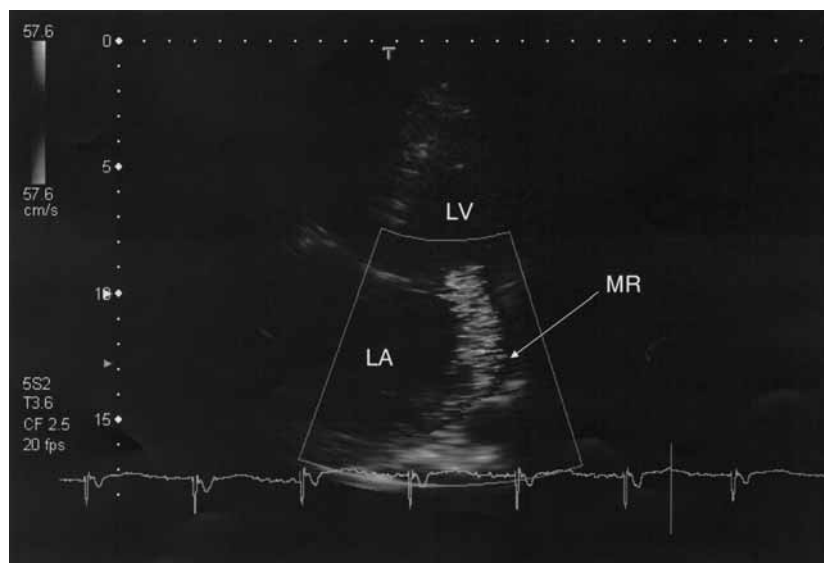


Fig. 1. Pre-operative echocardiography showing MR. MR was detected by echocardiography. The grade was “severe” by determination of the jet distance, but the width of the regurgitant jet was relatively narrow. The anterior mitral leaflet was slightly prolapsed. LA, left atrium; LV, left ventricle; MR, mitral regurgitation.

LV wall motion and increased MR (grade 2): ejection fraction (EF) dropped from 54% in 2003 to 37% in 2004. Central venous pressure (CVP) was elevated to 16 mmHg (previously 12 mmHg). In addition, an arteriovenous fistula was detected by echo at the right femoral artery and vein due to the previous catheter examination: the shunt was 5×10 mm in size and its peak flow was at 2.4 m/s. Consequently, it was considered that the cause of PLE was congestive heart failure with reduced LV wall motion, MR and elevated CVP indicating surgery.

The right groin was opened and the arteriovenous fistula between the right femoral artery and vein was ligated. Median sternotomy was performed. CPB was initiated with aortic, pulmonary artery and right femoral vein cannulation. After the aortic cross-clamp and myocardial protection, the right side of the LA was opened. The mitral annulus was dilated, and the anterior mitral leaflet was slightly prolapsed but not degenerative. Mitral annuloplasty was carried out using a 30 mm Carpentier ring. A water test showed no MR. The LA was closed and a 5-mm-diameter fenestration was created by directly connecting the TCPC conduit and LA, and he was smoothly weaned off CPB. CPB and aortic cross-clamp time were 107 and 66 minutes, respectively.

His symptoms improved after the operation. His body weight decreased by 12.3 kg (from 67.8 kg at pre-operation to 55.5 kg at post-operation). Post-operative CVP was 10 mmHg, and the levels of total protein and albumin returned to the normal range in 2 weeks (Table 1). He was discharged on the 39th post-operative day in a stable condition. He has been followed up monthly in the out-patient clinic without

recurrence of PLE for 1 year (Table 1). Echocardiography at 1 year showed only trivial MR although EF was decreased from 54.9% to 36.5%.

Comment

PLE is a serious clinical condition that occurs in 2.5-11% of patients who undergo Fontan operation, causing high mortality (50% at 5 years).^{1,2)} Causes and mechanisms of PLE are unclear, although it is speculated that the elevated systemic venous pressures associated with the Fontan circulation cause the intestinal protein loss by increased pressures in the enteric lymphatic system.¹⁾ Other risk factors for PLE are longer CPB time, single RV anatomy,²⁾ infection, inflammation,⁵⁾ and low cardiac output resulting in increased mesenteric vascular resistance.⁶⁾ Extracardiac TCPC can achieve superior hydrodynamic efficiency and reduction of arrhythmias compared to other types of Fontan procedures.⁷⁾

RV exclusion procedure was first described by our group^{3,4)} and this patient had a good quality of life for 7 years after the first operation. Possible causes of PLE in this case might be congestive heart failure with LV dysfunction and elevated systemic venous pressure. Furthermore, it was speculated that the possible causes of LV dysfunction were increased MR, delayed influence of ARVD to LV, volume overload for right heart system due to the arteriovenous shunt at femoral site, and/or other unknown factors. PLE is often resistant to medical treatment (diuretics, heparin, steroids, etc.), diet (low fat and high protein), a non-surgical interventional approach, and

Table 1. Results of surgical operation for protein-losing enteropathy

		Pre-operation	Post-operation	
			Pre-discharge	1 year post-operation
Lab-data	Total protein (g/dl)	4.5	7.1	6.6
	Albumin (g/dl)	2.8	4.3	4.1
Echocardiography	LVDd (mm)	56	52	52
	LVDs (mm)	41	37	42
	Ejection fraction (%)	51.7	54.9	36.5
	Fractioning shortening (%)	26.8	28.7	17.6
	Mitral regurgitation	Severe*	Trivial	Trivial
Cardiac catheter examination	Ejection fraction (%)	37.0	-	-
	Cardiac output (L/min)	2.49	-	-
	Cardiac index (L/min/m ²)	1.39	-	-
Central venous pressure (mmHg)		16	10	-
SaO ₂ at room air (%)		94-96	80-84	84-85
Symptoms	Diarrhea	(+)	(-)	(-)
	Edema	(+)	(-)	(-)

*, Mitral regurgitation was grade 2 at catheter examination.

LVDd, left ventricular end-diastolic diameter; LVDs, left ventricular end-systolic diameter.

surgery. It is very difficult to decide the timing of surgery for PLE. However, if all the other treatments are not effective against serious PLE, surgery needs to be considered as an option.

It was unclear that surgical procedure was effective for PLE in this patient; fenestration, mitral valve repair, and/or ligation of arteriovenous fistula. Fenestration for this case could be controversial because it might not be necessary if the main cause of PLE was MR. In addition, if MR recurs post-operatively, L-R shunt will increase and CVP will elevate again. However, in this case, we considered that the main causes of PLE were congestive heart failure and volume overload of the right heart system. As leaflets of mitral valve were almost normal, we thought the chance of MR recurrence was small and mitral valve replacement was not needed. It was expected that the fenestration reduced the right heart overload. In fact, congestive heart failure was improved and CVP was decreased. In out-patient clinic, PLE has not recurred with only trivial MR, while, the EF has deteriorated. The cause of decreased cardiac function was unclear, but it might be myocardial degeneration like cardiac myopathy, hypoxia, or delayed influence of ARVD to LV. Further careful follow-up will be needed using an echocardiography.

RV exclusion procedure for this case is different from Fontan procedures for congenital heart disease in children from the points of view of anatomical background and hemodynamical change between pre- and post-operation.

However, surgery successfully treated PLE which was resistant to medical treatment.

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