Protein losing enteropathy secondary to a pulmonary artery stent

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ABSTRACT

A 2-year-old patient with hypoplastic left heart syndrome presented 6 months following Fontan completion with protein-losing enteropathy (PLE). He had undergone stent implantation in the left pulmonary artery after the Norwood procedure, followed by redilation of the stent prior to Fontan completion. Combined bronchoscopic and catheterization studies during spontaneous breathing confirmed left bronchial stenosis behind the stent, and diastolic systemic ventricular pressure during expiration of 25 mm Hg. We postulate that the stent acts as a valve, against which the patient generates high expiratory pressures, which are reflected in the ventricular diastolic pressure. This may be the cause of PLE.

Keywords: Bronchial compression, stents, protein losing enteropathy

INTRODUCTION

Protein-losing enteropathy is a feared late complication of all forms of the Fontan operation. The etiology of this disease is unclear, but it may be associated with increased systemic venous pressure, systemic ventricular dysfunction or AV valve regurgitation, or mechanical obstructions affecting the Fontan circuit. We report a new putative cause of PLE in a young patient.

A term neonate with hypoplastic left heart syndrome underwent a Norwood type of palliation in the neonatal period, followed by a bidirectional superior cavopulmonary shunt at 4 months of age. The postoperative course was complicated by recurrent pleural effusions. Cardiac catheterization demonstrated a small left pulmonary artery which was treated by stent implantation [Figure 1a]. At 2 years of age, the stent was redilated to 10mm diameter, to match the diameter of the right pulmonary artery [Figure 1b], followed by Fontan completion using an extracardiac conduit. In the postoperative period, the patient was diagnosed to have a stenosis of the left bronchus, but could be weaned successfully from the ventilator. Six months later, he presented with protein losing enteropathy (PLE). A combined cardiac catheterization and bronchoscopic

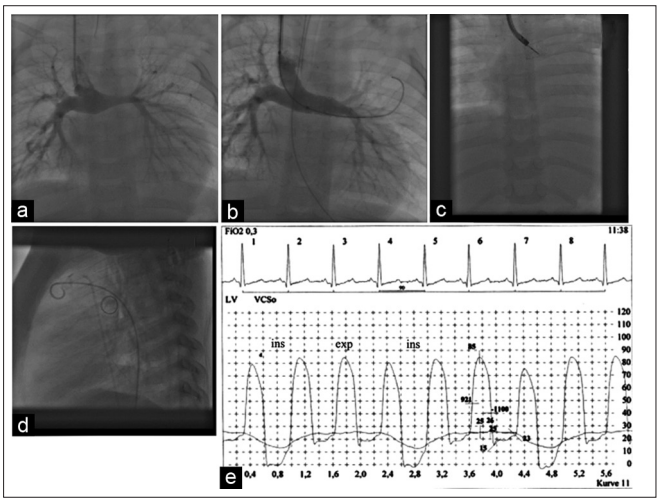

Figure 1: (a) Pulmonary artery angiography after the superior cavopulmonary shunt, showing a small left pulmonary artery. (b) Left pulmonary angiography following redilation of stent to 10 mm diameter, prior to Fontan completion. (c) Bronchoscopy after Fontan completion. The thin metal probe does not pass beyond the stent in the left pulmonary artery. There was a >70% stenosis of the left main bronchus at this site. (d) Lateral radiographic projection (the stent in the left pulmonary artery is seen end on). The stent compresses the bronchus directly behind it. There is an angiographic catheter introduced retrogradely to the neo-ascending aorta. (e) Simultaneous systemic ventricular and pulmonary arterial pressure traces during spontaneous breathing. During inspiration (beats 1, 4, 7) the ventricular diastolic pressure drops to normal physiological values, with a mean pulmonary artery pressure of 12 mm Hg. During expiration (beats 2, 3 and 5, 6 and 8), the ventricular diastolic pressure increases to 25 mm Hg, with a concomitant increase in the mean pulmonary artery pressure (scale given on the right hand side of the diagram, in mm Hg)

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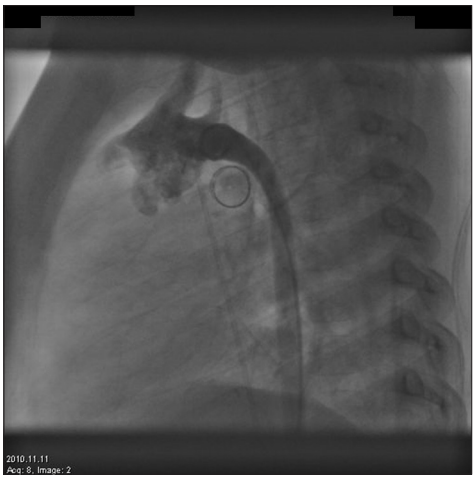

Figure 2: Aortic angiography showing no bronchial compression by the aorta

study were performed, with the patient breathing spontaneously. There were no stenoses in the Fontan pathways. The left bronchus was severely obstructed (>70%), directly behind the stent in the left pulmonary artery [Figure 1c and 1d]. Simultaneous systemic ventricular and pulmonary arterial pressure traces demonstrated a rise in the ventricular diastolic pressure during expiration to 25mm Hg. During inspiration, the end diastolic pressure normalised, with a reduction in the mean pulmonary artery pressure to normal values for the Fontan physiology [Figure 1e]. Bronchial compression by aorta was also ruled out [Figure 2]. We postulate that the stent acts as a valve, with the child generating high intrathoracic expiratory pressure against the stent, which in turn is reflected in the behaviour of the ventricular diastolic pressure. The intermittent elevation in systemic venous pressure is probably causative for PLE.

Follow-up

The patient is currently under a trial of conservative management (oral corticosteroid therapy and albumin replacement therapy), and is at home. Reoperation (removal of the stent) and patch-plasty of the LPA is under consideration.

DISCUSSION

In current cohorts, the Fontan operation is associated with improved outcomes and increasing short and long-term survival.[1,2] It may be expected that a significant proportion of patients will reach their teens and young adult life. Factors accounting for late attrition include systemic ventricular dysfunction, AV valve regurgitation, changes in the pulmonary vascular bed, late arrhythmias and PLE. Previous studies have demonstrated that PLE is associated with a significantly worse outcome,[3,4] and every effort should be made to manage these patients aggressively, using a combination of optimal pharmacological therapy and surgical or catheter based interventions to relieve mechanical problems affecting the Fontan circuit.[2,5-7] In the patient presented here, we postulate a new mechanical problem as being causative for PLE, with intermittent sharp increases in intrathoracic pressure accounting for elevation of the systemic venous pressure, and concomitant intestinal protein loss.

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