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Brief Report

Intra-aortic balloon counterpulsation in a patient with the failing Fontan circulation

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Abstract We report a 23-year-old patient undergoing functionally univentricular repair who developed cardiac failure due primarily to systemic ventricular dysfunction. The failing Fontan circulation was successfully re-established using intra-aortic balloon counterpulsation. Aortic counterpulsation facilitates recovery of systemic ventricular function, and appears to be a reasonable alternative in select instances of cardiac failure in patients with the Fontan circulation.

Keywords: Functionally univentricular repair; mechanical circulatory assistance; heart failure

As far as we are aware, thus far there has been limited use of intra-aortic balloon counterpulsation to support the failing Fontan circulation. We discovered 21 instances of such treatment, albeit with almost two-thirds dying.[1–5] In this report, we describe successful use of intra-aortic balloon counterpulsation in an adult patient with a failing Fontan circulation.

Case report

A 23-year-old male patient was referred to us with central cyanosis since 4-months of age, and dyspnoea on exertion. Clinical examination, chest roentgenogram, cardiac catheterization and angiocardiography confirmed the diagnosis of a functionally univentricular heart of indeterminate morphology, pulmonary stenosis, and a left-sided aorta. The systemic ventricular pressure was 110/0–10 millimetres of mercury, and there was global ventricular hypokinesia. The pulmonary arteries were confluent, the mean pulmonary artery pressure was 11 millimetres of mercury, and the pulmonary vascular resistance was 0.8 woods units per square metre. The patient underwent construction of a lateral tunnel total cavopulmonary connection, using a polytetrafluoroethylene patch (WL Gore and Associates, Flagstaff, Arizona, USA) with a 5 millimetre fensestration. The superior cavopulmonary connection was carried out on a beating, perfused heart under cardiopulmonary bypass. Antegrade cold blood cardioplegia was used for myocardial protection during completion of the Fontan connection. The periods of aortic cross-clamping, and bypass, were 48 minutes and 108 minutes, respectively. Postoperatively, he had stable haemodynamics on dopamine and sodium nitroprusside, with a mean left atrial pressure and central venous pressure between 6 and 8 millimetres of mercury. Eighteen hours after surgery, he developed signs and symptoms of low cardiac output. Despite optimal inotropic support using dopamine and dobutamine with expansion of the circulating volume, no haemodynamic improvement was observed. The mean left atrial and central venous pressures were 16 and 18 millimetres of mercury, respectively. Pharmacological afterload reduction was limited because of systemic hypotension. Cross-sectional echocardiography demonstrated wide and unrestrictive anastomoses, and unobstructed venous flow to both pulmonary arteries. The solitary ventricle, however, was grossly hypokinetic, with an ejection fraction of 0.38.

Considering the overall clinical picture, a trial of intra-aortic balloon counterpulsation was planned

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prior to take-down of the Fontan circulation. The thermodilution cardiac index was only 1.5, and mixed venous oxygen saturation was 24%. We used the Datascope system 97e pumping console (Datascope Corporation Cardiac Assist Division, New Jersey, USA) with a true sheathless 9.5 French, 40 cubic centimetre intra-aortic balloon catheter. The balloon was timed with the R-wave of electrocardiogram. It produced an immediate improvement in his blood pressure with suprasystolic diastolic augmentation. After 6-hours on balloon support, his cardiac index had increased to 2.8, and mixed venous oxygen saturation had risen to 68%. After 96 hours, he was successfully weaned from balloon support, and maintained stable haemodynamics. Doppler echocardiogram revealed normal systemic ventricular function, with a functioning fenestration.

Discussion

Therapeutic options for the failing Fontan circuit include medical manipulations, such as the use of nitric oxide, cardiac pacing, devices to improve venous return, such as intermittent compression of the abdomen or lower body, revision of Fontan connection, conversion to a hemi-Fontan circulation, mechanical circulatory assistance, and finally, orthotopic cardiac transplantation.[1–6] Supraventricular arrhythmias, systemic ventricular dysfunction, structural problems, and collateral circulation are among the major causes of failure. Because of this heterogeneity, the options for management need to be individualized.[6]

Take-down is usually beneficial if the aetiology is obstruction within the Fontan circulation, or increased pulmonary vascular resistance unresponsive to medical manipulations.[6] If the Fontan circulation cannot be sustained by the currently available medical treatment, with the pressure more than 16 millimetres of mercury, and mean left atrial pressure more than 12 to 14 millimetres of mercury, with a diagnosis of systemic ventricular dysfunction, the next strategy may be to assist the failing Fontan circulation by mechanical circulatory assistance. Take-down does not usually benefit this subgroup of patients.[1–6]

Extracorporeal membrane oxygenation and left ventricular assist devices are the most prevalent means of mechanical circulatory assistance in children. Although the use of balloon counterpulsation is universal in adults with acute left ventricular dysfunction after myocardial infarction or cardiac surgery, its use in patients with a failing Fontan circulation remains sporadic.[1–6] Use of balloon pumping for supporting the failing myocardium also remains limited in children, with variable success.[2–6] This is due to technical difficulty in inserting balloons in infants or small children, along with the availability of such balloons, and inability to track rapid heart rates and narrow pulse pressures of children in shock.[1–5] Additionally, complications like ischaemia of the limbs, renal failure, and mesenteric ischaemia are greater for smaller children because of inappropriate lengths of the balloons. Recent reports of specially designed balloons, nonetheless, have decreased the incidence of these complications.[1–5]

Early concerns of achieving effective counterpulsation in the highly elastic and distensible aorta of young children have proved unfounded.[4,5] The major concern that has hindered the widespread use of intra-aortic balloons in children, in contradistinction to adults, is that children are less likely to have preserved right ventricular and pulmonary function, and may not be supportable with the intra-aortic balloon. Physiologic differences between children and adults also necessitate more accurate timing of the counterpulsation.[5] Previous reports have demonstrated large errors in timing using electrocardiographic or arterial tracings.[5] The echocardiographic technique offers a more accurate method of timing the counterpulsation relative to opening and closing of the aortic valve.[7]

The timing and indications for deployment of the balloon is a matter of judgment, and may indeed be difficult. In patients who cannot be weaned from cardiopulmonary bypass, or in those who suddenly deteriorate after a good surgical repair, the decision to initiate balloon counterpulsation is relatively straightforward. For those with progressive deterioration of ventricular function, the decision can be more difficult.

Our search of the literature revealed 21 cases of patients with a failing Fontan circulation treated with intra-aortic balloon support. Only one-third of these, 8 of 21, survived (Table 1).[1–5] The current results from the last decade with balloon counterpulsation in infants and children undergoing cardiac surgery for complex congenital heart defects report an overall survival of 62%. These results approach the survival rate seen in adult series, of nearly 80%.[5,8]

The advantage of balloon counterpulsation over the use of left atrial-aortic assist devices is the ease of application.[1,5,8] Moreover, in the Fontan circulation, balloon pumping facilitates recovery of left ventricular function by decreasing left ventricular end-diastolic and left atrial pressure, thus increasing the driving force for the pulmonary circulation. Other assist devices, like axial flow pumps, have also been tried in experimental setting.[8]

We advocate caution against the widespread use of balloon pumping after the modified Fontan procedure. Clearly, there must be an exhaustive search for residual obstruction or other defects. Timing of initiation of support remains difficult, since its use as an absolute 'last resort' decreases the possibility of success. Randomized studies should be performed to define

Table 1. List of the 21 published cases of patients with a failing Fontan circulation treated with intra-aortic balloon counterpulsation.[1–5]

Authors	No. of Fontan	IABP death	Survivors
delNido PT et al. (1988)	9	8 (88.9%)	1 (11.1%)
Nawa K et al. (1988)	3	2 (66.6%)	1 (33.3%)
Park JK et al. (1993)	2	2 (100%)	–
Akomea-Agyin C et al. (1999)	5	1 (20%)	4 (80%)
Pinkney KA et al. (2002)	2	–	2 (100%)
Total	21	13 (62%)	8 (38%)

specific indications, proper time of intervention, and factors that can predict a successful outcome.

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