Modified Blalock-Taussig Shunt and Levosimendan for Left Ventricular Preparation in a Child with Transposition of Great Arteries and Regressed Ventricle Undergoing Rapid 2 Stage Arterial Switch Operation

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Abstract

Rapid two-stage arterial switch operation (ASO) is very relevant as many patients of transposition of great arteries (TGA) present late to the hospital when primary switch either is not possible or carries a high risk of morbidity and mortality. Hence, other means apart from the traditional methods of left ventricle preparedness should be tried to help this category of patients, who are to undergo rapid two-stage ASO. We successfully used levosimendan and continuous positive airway pressure after 1st stage operation in a patient with dTGA and regressed ventricle, which helped in left ventricular preparedness, and the child underwent rapid two-stage ASO uneventfully.

Keywords: *Blalock-taussig shunt, continuous positive airway pressure, left ventricular preparedness, levosimendan, rapid two-stage arterial switch operation*

Introduction

Arterial switch operation (ASO) has transformed the way patients of transposition of great arteries (TGA) were being managed. Unlike mustard and senning operations, which provided only physiological correction, ASO provided with both anatomical and physiological correction. Results of ASO performed in patients within first 4 weeks of life have been encouraging. However, beyond this period, there is a progressive decline in left ventricular (LV) function, as it is supporting low resistance pulmonary circulation. This is more troublesome when patients have an intact ventricular septum (IVS). In patients having TGA with ventricular septal defect, volume overload and high pulmonary artery (PA) pressure, help in preventing regression of LV myocardial mass, is notably absent in those having TGA with IVS. Hence, left ventricle is often regressed in these patients presenting late after neonatal period and not capable of supporting systemic circulation after ASO. They may be the candidates requiring ASO, but results in this subset of patients are unclear. Hence, two-stage ASO is recommended in late presenters in whom PA banding along with Blalock-Taussig (BT) shunt is done followed by ASO as the second step. We present a unique case where debanding of PA had to be done due to fall in oxyhemoglobin saturation (SpO_2) and arterial oxygen tension (PaO_2). To hasten the process of LV preparation, mechanical support of continuous positive airway pressure (CPAP) ventilation and pharmacological support with levosimendan, a calcium ion sensitizer, were started before second stage operation. The patient underwent successful ASO after 48 h with uneventful postoperative recovery.

Case Report

A 1-year-old girl weighing 6.5 kg was diagnosed to have TGA with IVS and regressed banana-shaped left ventricle on echocardiography. LV end diastolic dimension (LVEDD) was 16 mm, and posterior wall (PW) thickness was 3 mm [Figure 1]. Normal LV dimensions, i.e., LVEDD and PW thickness for this child would be 20 and 4.5 mm, respectively. A rapid two-stage ASO was planned for her illness. PA banding and a central shunt (4 mm) was done as first stage operation. However, in postoperative period patient developed desaturation (SpO_2 <60% and PaO_2 <40 mmHg), and her hemodynamics deteriorated after few hours of surgery for

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which debanding of the PA was done. The intolerance to the PA band could be because of the tightness of the band and the relative LV-PA gradient. Relying on central shunt alone for LV preparedness and safety in proceeding for the 2nd stage operation was doubtful. We decided to use CPAP and levosemendan in this patient to achieve faster and effective LV preparedness.

The child was weaned from the controlled mode (pressure regulated volume control) of ventilation to CPAP mode within next 24 h after the PA band removal. For next 18–20 h, she remained intubated and was ventilating spontaneously, on CPAP mode at 10 cmH_2O of PEEP through the endotracheal tube (CPAP) till the 2nd stage surgery. She was sedated with injection clonidine 30 µg through Ryles tube every 6 hourly, and she tolerated CPAP well. She was also started on levosimendan infusion at a dose of 0.05 µg/kg/min. Serial echocardiograms showed increase in LVEDD (from 16 to 18 mm) PW thickness (from 3 to 4 mm) and improvement in LV shape (from banana shape to D shape to spherical shape) and function [Figures 2 and 3]. The 2nd stage surgery (shunt take down and ASO) was performed after 48 h. The levosimendan infusion was continued for next 24 h in the postoperative period after 2nd stage surgery. The child recovered uneventfully and was discharged from the hospital on 12th postoperative day.

Discussion

With progressive fall in pulmonary arterial pressure after the birth, the afterload imposed upon the LV decreases, leading to a regression of LV mass. This restricts the safe period for ASO to 1 month.[1] For this subset of patients, LV preparedness in the form of PA banding and a aortopulmonary shunt, followed 5 months later by ASO was first reported by Yacoub *et al*. in 1977.[2] There was a problem with this approach, in the form of proximal main PA dilatation, neo-aortic valve regurgitation and the addition of prosthesis for reconstruction of neo-PA. These disadvantages were ameliorated in 1989, when rapid two-stage ASO was proposed, because of short duration between the two procedures.[3] A two stage ASO is well proven and documented procedure in these patients. Rapid LV training occurs by LV remodeling caused by increased pressure load, volume load, and wall shear stress. BT shunt causes a volume overload to LV, helping in LV preparedness. BT shunt in itself can increase PA pressures and help in preparedness. Aoshima *et al*.,[4] have reported conflicting report regarding LV preparedness using BT shunt alone. PA banding increases the afterload on the left ventricle, thus aid in its preparedness before ASO, but unfortunately, our patient did not tolerate the PA band and had to be removed.

Levosimendan helped the systemic ventricle to cope with the increased cardiac output returning to systemic ventricle. Furthermore, it helped the pulmonary ventricle by helping eject against systemic pressures at the level of PA due to BT shunt. Levosimendan is a calcium channel

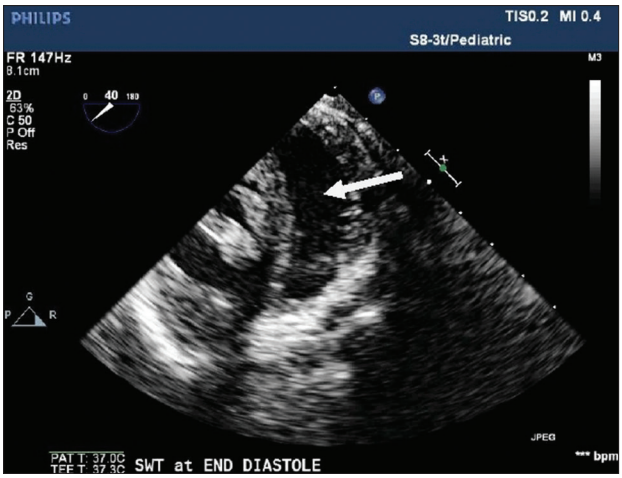

Figure 1: Transesophageal echo image showing typical "banana shaped" left ventricular cavity, indicated by solid white arrow (preoperatively during 1st stage operation)

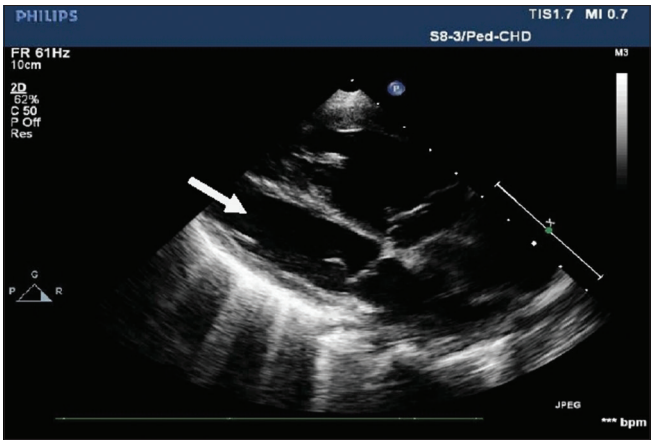

Figure 2: Transthoracic echo image showing D-shaped left ventricular cavity, indicated by solid white arrow (24 h after starting the patient on continuous positive airway pressure ventilation and levosimendan)

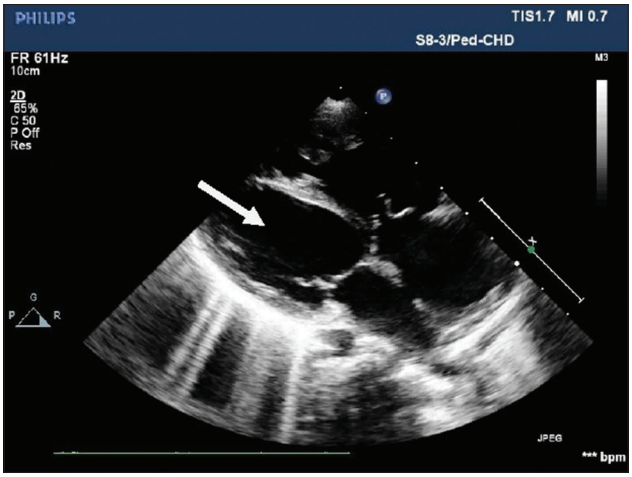

Figure 3: Transthoracic echo image showing spherical left ventricular cavity, indicated by solid white arrow (48 h after starting the patient on continuous positive airway pressure ventilation and levosimendan)

sensitizer which acts as an inodilator, like dobutamine and milrinone, but in contrast to them it does not cause much increase myocardial oxygen consumption; on the contrary, it has coronary vasodilatory properties. It thus increases cardiac contractility by binding to cardiac troponin C in calcium-dependent manner and stabilizes troponin C. This causes actin–myosin cross bridges without increasing intracellular concentration of free calcium. Levosimendan exerts its action in systole, and during diastole, desensitization is noted due to a plunge in the calcium concentration; therefore, ventricular relaxation is not hampered, and cardiac filling is optimum. Moreover, the metabolites of levosimendan R-1908 are very long acting, and its effect lasts for 7–10 days,[5] so it continues to decrease the LV afterload after ASO and prevents LV failure. Moreover, also the long half-lives of its metabolites allow time for the oral medications like angiotensin converting enzyme inhibitors to take over the unloading effect on LV. In a study by Namachivayam *et al*.,[6] Levosimendan was found to be absolutely safe in infants and children and allowed substantial reduction in catecholamine infusions with objective improvement in myocardial performance in children with acute heart failure. Lechner *et al*.[7] also reported the successful use of levosimendan in a premature male twin with TGA.

CPAP up to 10 cm H_2O decreases the heart rate and LV intramural pressure (during systole) thereby decreasing the systolic wall stress and myocardial oxygen consumption. CPAP has been shown to be helpful in congestive heart failure in many previous studies.[8] The mechanism involved appears to be related to CPAP-induced increases in intrathoracic pressure and afterload to the right ventricle (in case of TGA it is the afterload to LV). Moreover, also CPAP reduces cardiac preload, by impeding cardiac filling. CPAP was found to increase exercise tolerance in patients with stable congestive heart failure in a study by Chermont *et al*.[9] We in our case tried the CPAP along with the levosimendan to see if those can effectively train LV so that it can handle the afterload after ASO. It is difficult to say that such nonsurgical interventions can essentially substitute for the conventional approach for rapid two-stage ASO. However, from our case, it is observed that, in addition to BT shunt, application of these methods may hasten the LV preparedness and increases the safety margin for early 2nd stage surgery and also these approaches may facilitate a smoother recovery of these patients in the postoperative period. Long-term follow-up studies and randomized controlled trials are needed to validate this intervention. Many people have observed that over a short period of PA banding can prepare the LV for 2nd stage ASO. The same could have happened in our case too, which we consider as a limitation in this case report.

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Conflicts of interest

There are no conflicts of interest.

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