Ductal Stenting to Retrain the Involuted Left Ventricle in a Late Presenter Infant With Transposition of the Great Arteries, Intact Interventricular Septum

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Abstract

INTRODUCTION: Ductal stenting can be a nonsurgical option and less morbid method to provide pressure and volume overload to the regressing left ventricle in late presenters of transposition of the great arteries, intact ventricular septum (TGA-IVS), prior to arterial switch operation (ASO).

CASE PRESENTATION: This is a case of an infant diagnosed with TGA-IVS beyond the neonatal period who underwent balloon atrial septostomy and ductal stenting to retrain involuted left ventricle prior to definitive ASO.

DISCUSSION: Ductal stenting is an alternative option in late presenters of transposition of the great arteries with involuted left ventricle. It provides volume loading and, to a lesser extent, pressure loading leading to left ventricular hypertrophy. Also, presence of the stented duct allows improved oxygenation eliminating the need for aortopulmonary shunt. It can be a less morbid method of left ventricular training because it avoids hemodynamic stress, pulmonary artery distortion, and neoaortic valve regurgitation.

CONCLUSION: Ductal stenting in late presenters with TGA-IVS successfully retrains the left ventricle and achieves safe late ASO.

KEYWORDS: congenital heart disease, transposition of great arteries, PDA stenting, arterial switch operation

INTRODUCTION

Transposition of the great arteries (TGA) is the most common form of cyanotic congenital heart disease presenting in the newborn period and comprises approximately 5% of all congenital heart defects.[1]

Arterial switch operation (ASO) has become the preferred approach for repair of TGA, intact ventricular septum (TGA-IVS), for those younger than 3 weeks with good outcome and results.[2] However, patients with TGA-IVS presenting beyond the first few weeks of life with involuted left ventricle have been considered at high risk of ASO because of the failure of the left ventricle to cope with systemic work as pulmonary vascular resistance declines. Hence, alternative strategies, such as the atrial switch or rapid 2-stage arterial switch, have generally been used to retrain the deconditioned left ventricle. Pulmonary artery banding combined with a systemic–pulmonary shunt (usually a modified Blalock–Taussig anastomosis) as the preparatory stage of the 2-stage approach has the disadvantage of stormy postoperative course and high hospital costs.[3]

Ductal stenting can be a nonsurgical option to provide pressure and volume overload to the regressing left ventricle prior to ASO.[2] It has the advantage of being a less morbid method of left ventricular (LV) training because it avoids hemodynamic stress, pulmonary artery distortion, and neoaortic valve regurgitation.

This case report describes the case of an infant who was a late presenter of TGA-IVS with involuted left ventricle where we proceeded with ductal stenting to improve oxygenation and left ventricle training prior to ASO.

CASE PRESENTATION

The infant was born term to a 20-year-old G1 P0 mother who had unremarkable prenatal course. Patient was delivered term via normal spontaneous delivery in a local hospital and was discharged as a well baby after 24 hours of hospital stay. At 1 week of age, patient was noted to have cyanosis when crying; however, no consultation was done. The cyanosis was noted to be progressive; hence, at 3 weeks of age, consultation was done with a pediatrician, who referred the patient to a pediatric cardiologist. Diagnosis was made at 4 weeks of life, with congenital heart defect, dextro-TGA, IVS, restrictive patent foramen ovale, and closing patent ductus arteriosus (PDA) with systemic arterial oxygen saturation at 40%. Patient had emergency bedside balloon atrial septostomy using TMP-Ped balloon catheter size 10 × 20 mm under transthoracic echocardiography guidance. Interatrial defect post balloon measured 0.5 cm from 0.3 cm, and O_2 saturation improved to 80%. Patient was then referred to our institution for ASO.

Patient was seen in our institution at 5 weeks of life, and on physical examination, vital signs were the following: blood pressure 67/37 mm Hg, pulse rate 133 beats/min, respiratory rate 50 breaths/min, oxygen saturation of 68% on all extremities, weight of 3 kg, with adynamic precordium, apex beat at the fourth intercostal space left midclavicular line, S1 normal, S2 single, and grade 3/6 systolic ejection murmur at the left upper sternal border. Chest radiography revealed increased pulmonary vascularity and right ventricular prominence with narrowed pedicle. Electrocardiographic findings revealed sinus rhythm, extreme right axis deviation, and right ventricular hypertrophy.

Echocardiogram showed TGA-IVS, interatrial defect of 0.5 cm, and a closing ductus arteriosus. Also, a type III or “banana-shaped” LV geometry with interventricular septum bowing into the LV cavity, reduced LV mass, LV volume, and thinned free wall were noted (Table 1, Figure 3). Patient had desaturation as low as 60% despite prostaglandin infusion. In view of the poor saturation and involuted left ventricle, patient was deemed unfit for primary repair. Hence, a 2-stage approach was considered through stenting of the ductus to improve oxygenation and to retrain the left ventricle prior to definitive ASO.

The patient underwent hemodynamic studies (Table 2), which documented left-to-right ventricular pressure ratio of 0.55, suggestive of low LV pressure. Descending aortogram revealed a closing ductus arteriosus. The PDA was then accessed using a cut pigtail catheter with a coronary wire from the descending aorta to the pulmonary arteries. The coronary wire was then advanced through the PDA and was anchored to the peripheral pulmonary artery (Figure 1).

Table 1. Echocardiographic Characteristics Pre- and Post-LV Retraining

	Criteria for Arterial Switch Operation	Pre-LV Retraining (Aged 5 wk)	Post-LV Retraining (Post-PDA Stenting) (Aged 2 mo)
Indexed LV mass, g/m^2	≥50 g/m^2	38	65
LV end-diastolic volume, cm	>90% of normal (1.4–2.35)	1.5	2.28
Ventricular septal profile/LV geometry	Type I–II	Type III	Type II
LV posterior wall thickness, cm	≥4 mm	0.4	0.6
LV ejection fraction, %	≥0.5	78%	82%

LV=left ventricular; PDA=patent ductus arteriosus.

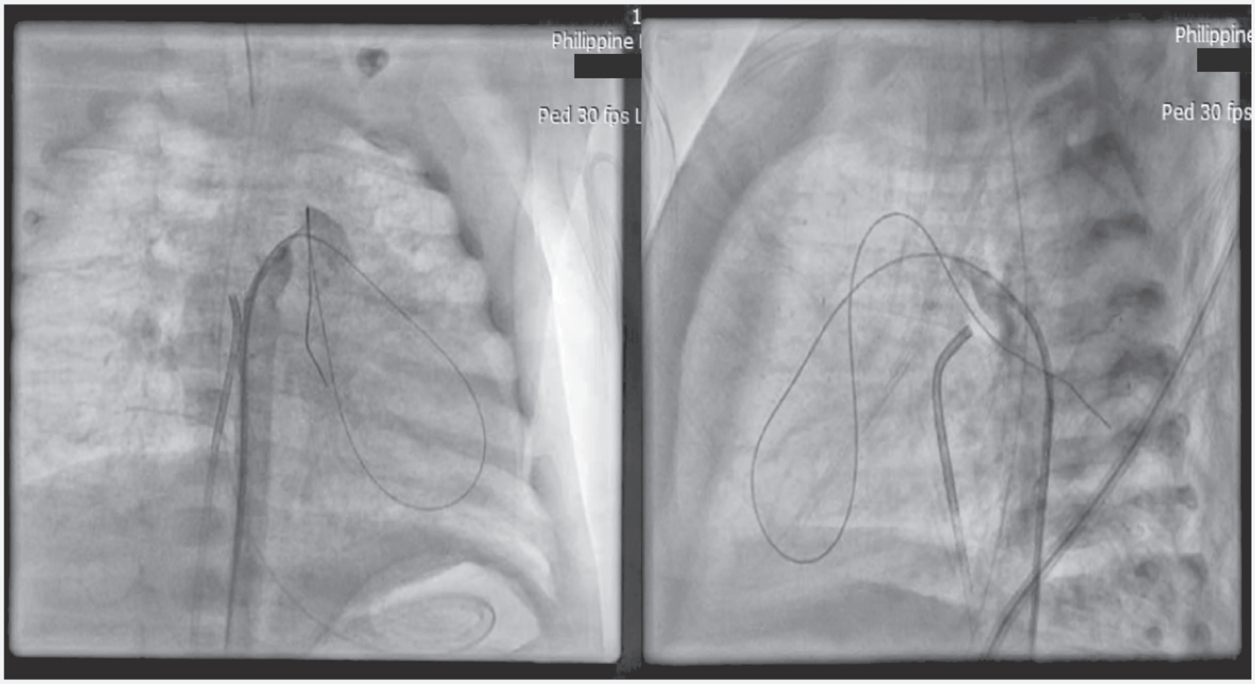

Figure 1. Descending aortogram in right anterior oblique and lateral views showing the closing PDA accessed by a coronary wire anchored into distal pulmonary artery

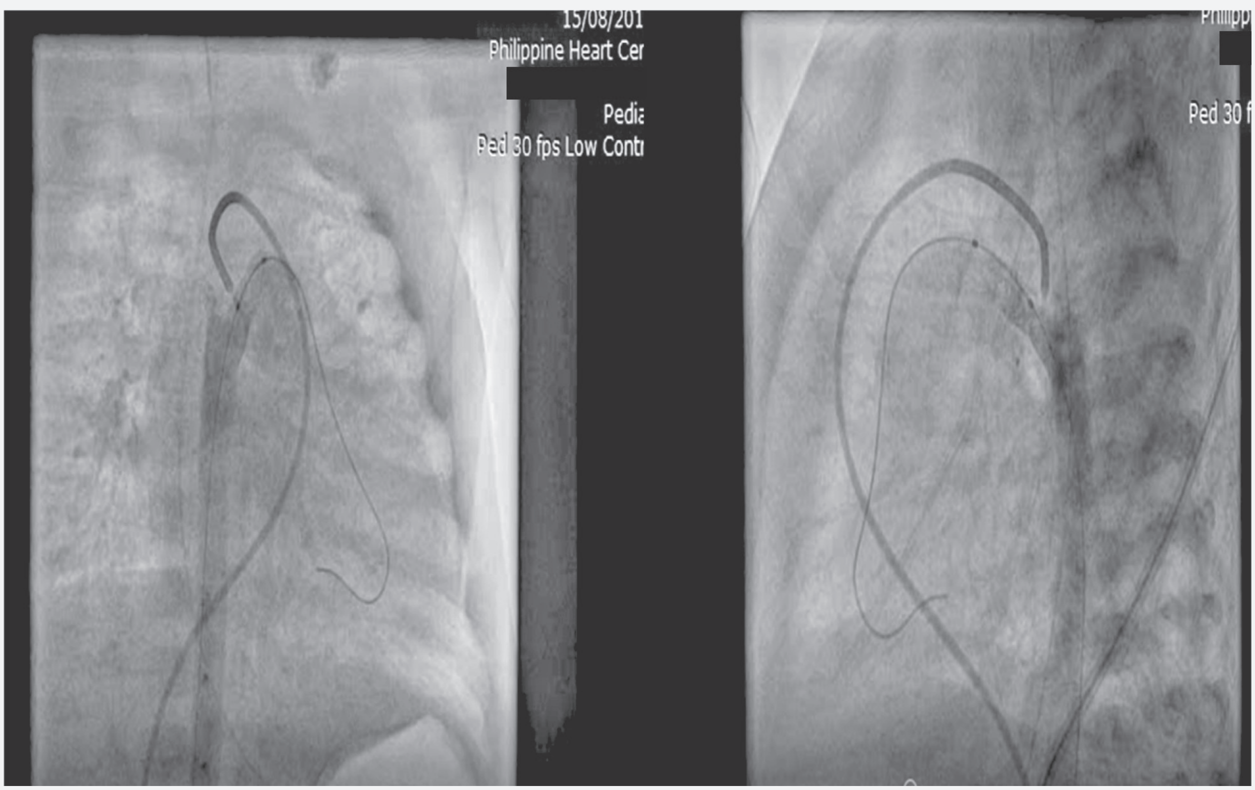

Figure 2. Descending aortogram in right anterior oblique and lateral views showing the ductal stent

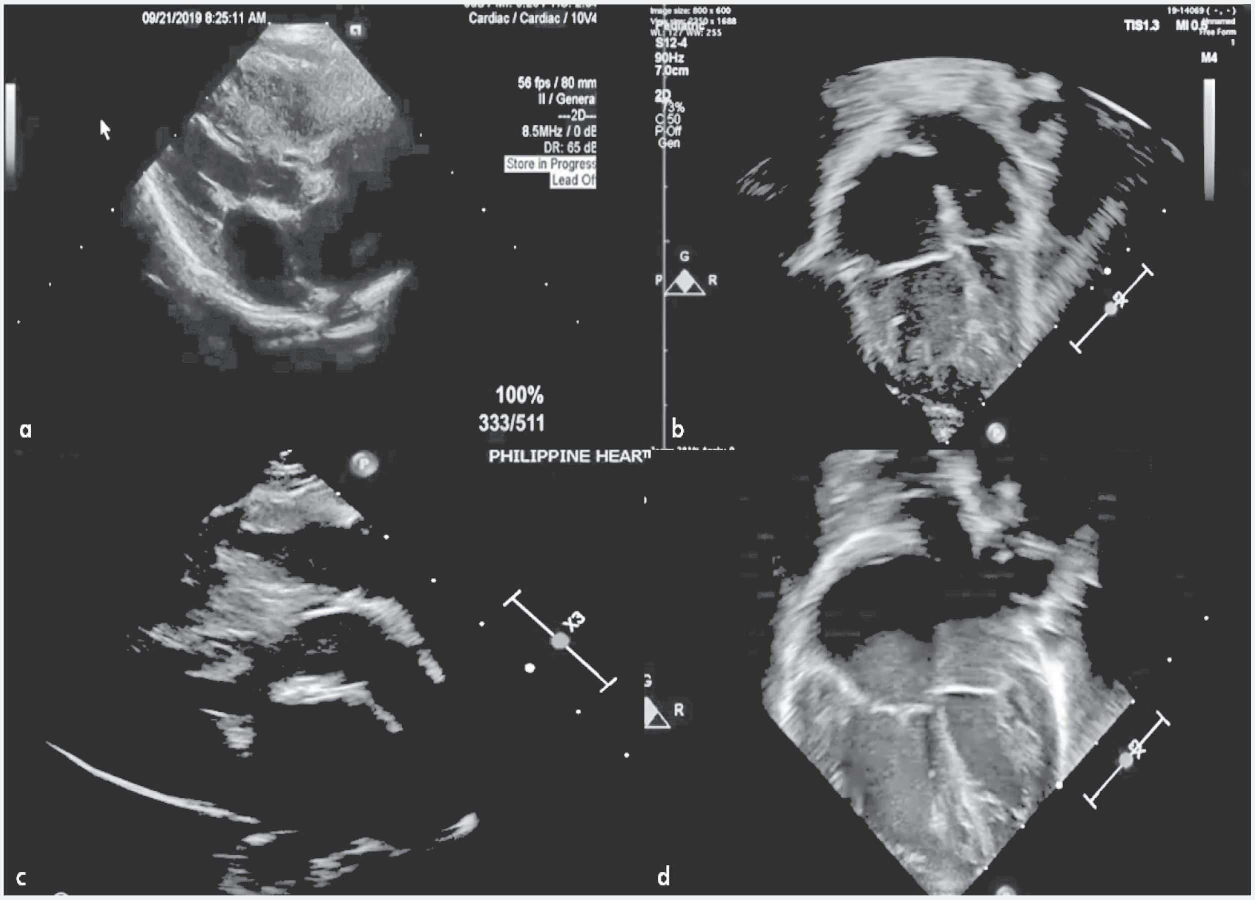

Figure 3. Echocardiographic views of the left ventricle showing comparable left ventricular wall thickness and volume before ductal stenting (A, B) and after 6 weeks of LV retraining (C, D)

Table 2. Hemodynamic Studies Pre- and Post-PDA Stenting

	Pre-PDA Stenting	Post-PDA Stenting
RV systolic pressure, mm Hg	79	74
LV systolic pressure, mm Hg	44	57
LV/RV pressure ratio	0.55	0.77
O_2 saturation	60%	90%

LV=left ventricular; PDA=patent ductus arteriosus; RV=right ventricular.

The cut pigtail catheter was then removed, then a Medtronic Resolute Integrity stent size 4 × 9 mm was introduced, and then the stent was dilated at the level of the PDA. The position was checked via hand shots of contrast while adjusting the position of the stent. Repeat angiography revealed adequately stented ductus arteriosus with no stent protrusion (Figure 2).

Patient's saturation improved up to 90%. Intravenous heparin was initiated and titrated accordingly, keeping activated partial thromboplastin time two times the normal value. Aspirin was overlapped with heparin infusion before heparin was weaned off. Patient was weaned off eventually from ventilation and inotropic support. Unfortunately, patient developed septicemia during admission, was treated with antibiotics, and was discharged and improved after 3 weeks of hospital stay.

After 6 weeks of LV retraining, repeat echocardiogram revealed an adequate left ventricle (Table 1, Figure 3). Patient was then referred to surgery for ASO. Patient underwent ASO at 3 months of age, and surgery was uneventful. Patient developed clinical sepsis during stay at the ward and was discharged and improved after 2 weeks of hospital stay. Echocardiogram before discharge demonstrated good ventricular functions and no neoaortic or neopulmonary stenosis or regurgitation.

DISCUSSION

Late presenters of TGA-IVS with involuted left ventricle have been considered at high risk of primary ASO because of failure of the deconditioned LV to cope with systemic work beyond the first few weeks of life. In our country, there are only a few government hospitals capable of performing congenital heart surgeries, and even at maximum capacity, early repair is not possible for many patients. The predominant reason for delayed surgery in the late switch group was late referral of patients because of socioeconomic and geographical factors. In our patient, diagnosis was made at 4 weeks of age presenting with desaturations; hence, a balloon atrial septostomy in a local hospital was performed to provide adequate mixing to improve oxygenation and then eventually referred to our institution for ASO. However, our patient was still desaturated, and echocardiogram revealed a "banana-shaped" LV geometry unsuitable for primary ASO. Thus, the patient was palliated via stenting of ductus arteriosus to improve oxygenation and retraining of the left ventricle.

Indications for LV training include a combination of the following noninvasive criteria: indexed LV mass of less than 35 g/m^2, age older than 3 weeks, ventricular septal profile, with a banana-like LV shape on two-dimensional echocardiograms, and absence of an LV outflow tract obstruction. Also, a left/right ventricular pressure ratio of less than 0.6 obtained by cardiac catheterization is an indication for a staged ASO.[2] All of these criteria were present in our patient, hence the LV retraining.

Pulmonary artery banding combined with a systemic–pulmonary shunt or modified Blalock–Taussig shunt has been used to retrain the LV. However, it is associated with stormy postoperative course, significant interstage morbidity and mortality, and higher procedural costs.[4]

Ductal stenting is an alternative option in late presenters of TGA with involuted left ventricle. It provides volume loading and, to a lesser extent, pressure loading leading to LV hypertrophy. Also, presence of the stented duct allows improved oxygenation, eliminating the need for aortopulmonary shunt. It can be a less morbid method of LV training because it avoids hemodynamic stress, pulmonary artery distortion, and neoaortic valve regurgitation.[2,4] Ductal stenting in TGA with IVS was first reported by Sivakumar et al[4] in 2006, where they described successful ductal stenting and subsequent ASO in two children aged 3 months. Also, in a study by Ota et al[5] in Malaysia, 10 patients underwent PDA stenting as LV retraining and as rescue stenting for severe arterial desaturation and had successful ASO.

Kothari et al[6] have described successful ductal stenting for TGA-IVS with regressed LV in five children aged 3 to 6 months. Ductal stenting resulted in LV preparedness within 7 to 14 days. In their study, our patients underwent successful uneventful arterial switch surgery, but one patient died of progressive sepsis after 14 days of stenting, even though the LV was prepared. In our patient, serial echocardiography was done, and LV preparedness was achieved, as shown in Table 1, 6 weeks after ductal stenting. This is similar to the study by Sivakumar et al,[4] where two patients who underwent ductal stenting for LV retraining showed LV preparedness after 24 to 28 days of PDA stenting.

The use of moderate-size (3.5 or 4 mm) coronary stents for ductal stenting was suggested to avoid postprocedure heart failure.[2] In a study by Kothari et al,[6] 3- to 3.5-mm coronary stents were used, where one of four patients required redilatation of the stent due to occlusion. This complication of reocclusion was found to be due to protrusion of the mucosal folds of the duct into the stent through the struts. All four patients, however, underwent successful ASO where stents were removed without any difficulty during surgery. In our patient, a 4-mm coronary stent was used with good flow noted until ASO was done.

In a study by Leong et al,[7] an atrial septal defect–to–interatrial septum length ratio of greater than 0.38 was associated with failed retraining. According to the European Association for Cardio-Thoracic Surgery and the Association for European Paediatric and Congenital Cardiology clinical guideline for the management of TGA-IVS,[2] it may be advisable not to pursue a wide ASD, to ensure adequate preload of the left ventricle. Creation of a small to moderate (4–5 mm) atrial septal communication should be considered. In our patient, interatrial communication was maintained at 0.5 cm, allowing adequate preload of the left ventricle.

Guidelines on optimal time interval between LV retraining and ASO have not yet been established, but the proposed criteria for a safe second-stage ASO include two-dimensional echocardiography and hemodynamic parameters.[2] In some studies, the median interval between stages is 10 days (range, 5 days to 6 weeks), as long as adequate LV mass and volume were achieved.[2,4,6] In developing countries such as ours, where children with complex congenital heart diseases need surgical intervention, socioeconomic and geographical factors are of great consideration as the primary causes of delay in diagnosis. Nevertheless, effective noninvasive alternative options and improvement in intraoperative and postoperative care have contributed to the improvement of the outcome of ASO in our institution.

CONCLUSION

Ductal stenting in late presenters with TGA-IVS is an effective alternative method to retrain a deconditioned left ventricle, given its noninvasive nature and reduced postprocedural morbidity.

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