**IMAGES** 

in PAEDIATRIC CARDIOLOGY

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### **Abstract**

Simple transposition of the great arteries (TGA) occurs in 0.2 per 1000 live births. The condition is surgically repaired in the neonatal period by the arterial switch procedure (ASO) sometimes preceded by an atrial septostomy. The ASO involves transecting the great arteries and relocating them to the appropriate ventriculo-arterial (VA) connection with attachment of the disconnected coronary arteries to the aorta. In the process, the attachment of the pulmonary artery to the right ventricle involves the Le Compte manoeuvre and to achieve this the pulmonary arteries must be fully mobilised and sometimes the main pulmonary artery may require patch augmentation as well. Nevertheless, pulmonary artery stenosis (PAS) is one of the potential problems with the ASO. However, with improved surgical techniques, this has dropped from around 15% in the 1980s to less than 3%. Apart from surgical revision when PAS occurs, there are interventional options which include angioplasty and/or stent insertion. The latter is preferred in small children and works well in around 60% but may require repeat procedures. In older patients or when angioplasty fails, stent insertion can be considered. These procedures may involve negotiating tight bends in order to reach the site of stenosis. The passage of non-premounted stents may be problematic in such situations, especially with longer stents and tighter bends as they tend to slip off balloon. We describe several techniques that may facilitate such interventions, and these were utilised in an adolescent patient who had had ASO for TGA in the neonatal period. These included manually giving the mounted stent a slight bend in order to help the balloon-stent assembly negotiate hairpin bends.

### Introduction

Transposition of the great arteries (TGA) occurs in 0.2 per 1000 live births,<sup>1</sup> and comprises approximately 5% of all congenital heart defects.<sup>2</sup> The condition is surgically repaired in the neonatal period by the arterial switch procedure (ASO). This involves transection of the proximal aorta above the aortic valve and moving the aorta posteriorly towards the left ventricle, along with dissection and relocation of the coronary arteries to the neoaorta. The pulmonary arteries are extensively dissected to minimise tension and the main pulmonary artery is transected and anastomosed to the right ventricular outflow. The original description of this by Jatene has been largely replaced by slinging the pulmonary arteries in front of and straddling the ascending aorta as described by LeCompte.<sup>3,4</sup>

This manoeuvre results in stretching of the pulmonary arteries which may result in pulmonary artery stenosis (PAS) if there is inadequate dissection or not enough vessel length.

PAS is a recognised complication of this manoeuvre that occurs in up to 10% of cases.<sup>5,6</sup> It has been speculated that this stenosis may be due to scar tissue formation at the anastomosis sites, inadequate somatic growth of the pulmonary artery and inadequate mobilization of both outflow tracts, with resultant tension at the anastomosis sites, as well as nonviable tissue at these sites;<sup>7</sup> it is likely to multi-factorial in many cases.

Surgical repair of such stenosis is possible but less invasive relief of obstruction is available by balloon angioplasty with or without stenting. Ballooning alone also carries a higher failure rate than stenting so stenting is employed in tight stenosis, but the age at presentation plays a role. In this respect, PAS either occurs soon after surgery and relatively rapidly or it can be a very slow process presenting later in life and associated with somatic growth.

Such procedures may involve negotiating tight bends in order to reach the site of stenosis. The passage of non-premounted stents may be problematic in such situations, especially with longer stents and tighter bends. Care must also be taken in order to avoid inflating stents very proximally as these may impinge on the coronary arteries. If there is any suspicion of this, balloon interrogation should be performed prior to stenting. We describe several techniques that may facilitate such interventions.

# **Case Presentation**

Our patient presented in the early neonatal period with simple TGA and underwent an uneventful ASO. Echocardiographic follow-up showed slowly progressive right ventricular outflow tract obstruction, as evidenced by tricuspid regurgitation gradients, but failed to elucidate the exact location of the obstruction.

Magnetic resonance imaging at fourteen years of age showed discrete narrowing of the right pulmonary artery (RPA successive orthogonal views – figures 1 and 2) with the narrowest orifice is 11x4 mm and Vmax 1.1 m/s. There was also long segment narrowing of the left pulmonary artery (LPA – figures 3 and 4) with a minimal orifice diameter of 2.5 mm. and Vmax 3.3 m/s. The main pulmonary artery was 9x18 mm

Figure 1: Successive Orthogonal views obtained by magnetic resonance imaging using a balanced steady-state free precession pulse sequence, showing discrete narrowing of the right pulmonary artery (arrowhead).

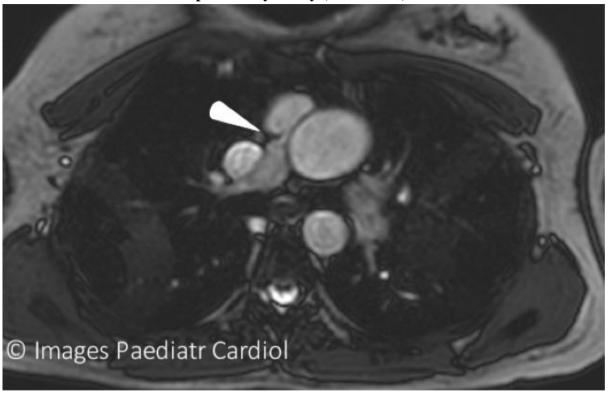


Figure 2: As figure 1



Figure 3: Orthogonal views obtained by magnetic resonance imaging showing narrowing long segment narrowing of the left pulmonary artery (arrowhead).

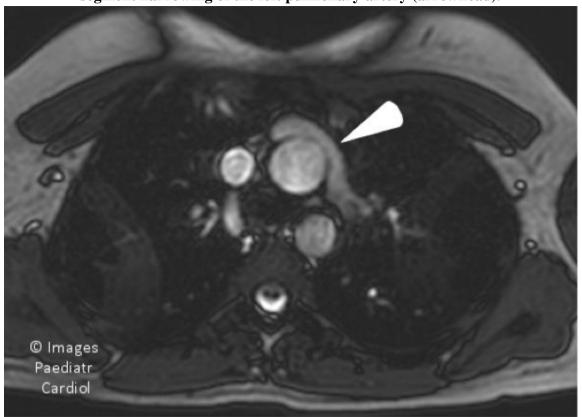
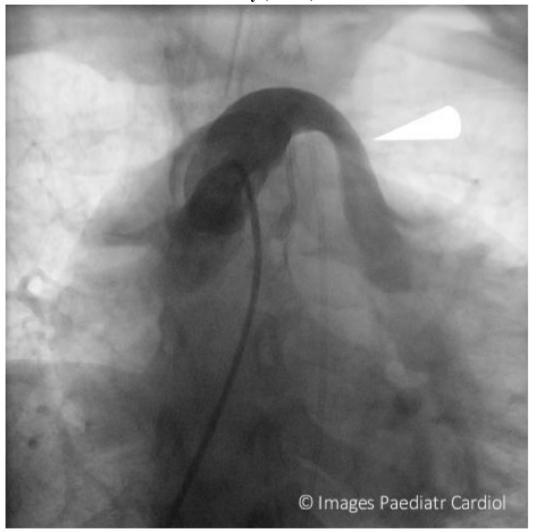


Figure 4: As figure 3.



She underwent cardiac catheterisation with the intention of balloon intervention and stenting as required. Angiography in steep anteroposterior steep cranial as well as steep caudal (45 degrees) angulations confirmed tight but discrete narrowing of the RPA origin and long segment narrowing of the proximal LPA (figure 5). An Amplatz superstar exchange 0,035" wire was used for the intervention.

Figure 5: Angiography showing narrowing long segment narrowing of the left pulmonary artery (arrow).



The RPA was ballooned with a 20 by 40mm Cristal balloon. A tight waist on the balloon was abolished with some recoil.

The LPA stenosis was refractory to balloon dilatation. After luminal measurements on fluoroscopic projections, an Andrastent XXL 35mm stent was hand-crimped on a 15mm by 40mm Balt Cristal balloon and an attempt was made to introduce this into the LPA through a 12F Mullins sheath. The Mullins sheath was successfully introduced into the LPA. However, when attempting to negotiate the stent and balloon over the apex of the LPA curve, the stent slipped back over the balloon. The balloon and stent were retrieved and the stent was recrimped, again by hand. The sheath was changed to 14F size but once again, the stent shifted on the balloon shaft. Balloon and stent were once again retrieved

and this time, after recrimping by hand, the stent was given a slight curve. After this manoeuvre, the stent traversed the bend without any problems and was deployed (figures 6 and 7).

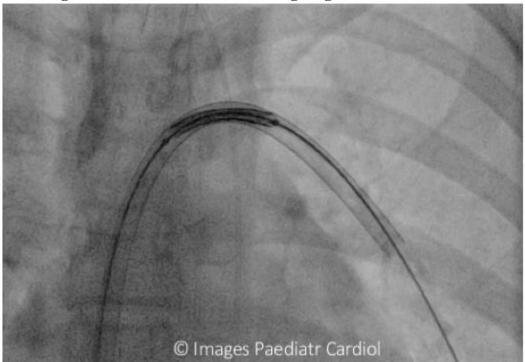
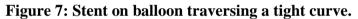
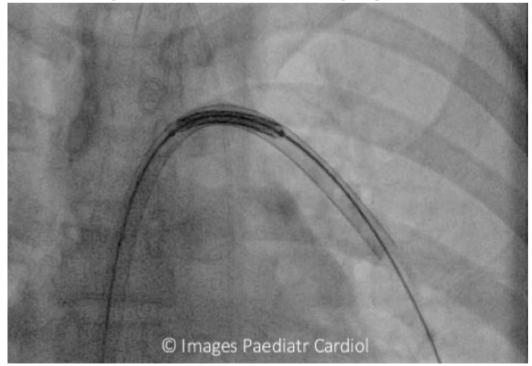


Figure 6: Stent on balloon traversing a tight curve into the LPA





The stent was successfully deployed (figure 8) with reduction of right heart pressures (table 1).

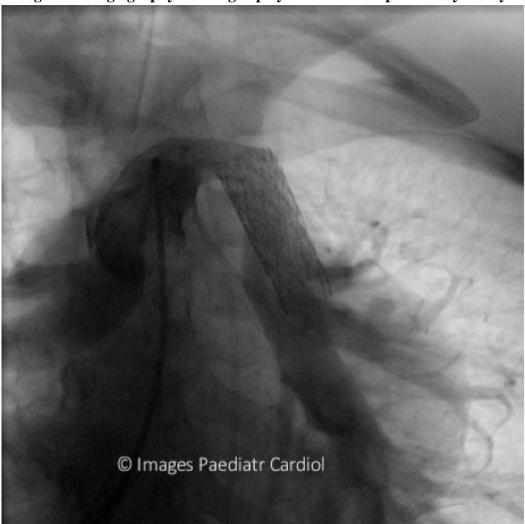


Figure 8: Angiography showing deployed stent in left pulmonary artery

**Table 1: Pressures during the procedure (mmHg)** 

Aorta	100/80	Throughout
Pre		Pullbacks
RV	45/7 mean 15	RPA to MPA: 14/8, mean 10 to 44/8 mean 20
		LPA to MPA: 20/12 mean 15 to 44/8 mean 20
Post		
MPA	30/4 mean 10	
LPA	23/5 mean 12	
RPA	15/7 mean 10	
RV	45/7 mean 15	

LV=left ventricle, RV=right ventricle, RPA=right pulmonary artery, LPA=left pulmonary artery

#### Discussion

We describe a case of bilateral branch pulmonary stenosis in an adolescent patient following the LeCompte manoeuvre carried out in infancy for ASO in a patient with TGA. Both stenoses were successfully treated percutaneously by balloon angioplasty and stenting, but the procedure was punctuated by challenges in negotiating a tight bend in the LPA. Steep cranial or caudal angiograms should be obtained in order to decide on the best view to image and balloon/stent pulmonary artery stenosis. In the case of non-premounted stents, for the negotiation of tight curves within long sheaths, a slight curve of the stent on the balloon may be useful to negotiate a tight curve, particularly if the stent is on a small or low profile balloon. But any such manoeuvres must be done cautiously so as to avoid damaging the balloon. An additional benefit of introducing a slight bend on the stent after mounting and hand-crimping is that this increases the stability of the stent on the balloon. This technique assumes that other steps would have already been taken, such as, the use of a stiff wire parked distally in a PA branch and upsizing the sheath to augment the lumen.

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