

Evolution of Approach to Right Ventricular Outflow Tract Stenting in Infants \leq 2Kgs

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Abstract

Surgical palliation or repair of symptomatic Tetralogy of Fallot in the neonatal period is associated with a relatively high mortality rate. Stenting of the right ventricular outflow tract is a newer procedure that has evolved to allow its performance in low birth weight neonates. In this case series, we describe the evolution of our approach to right ventricular outflow tract stent implantation in neonates weighing < 2 kg.

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Key Words

Congenital heart disease • Cardiac catheterisation • Tetralogy of Fallot • Low birth weight

Introduction

Surgical palliation or repair of symptomatic Tetralogy of Fallot (ToF) in the neonatal period is associated with a relatively high mortality rate. A large contemporary database of over 3,000 patients reports 6.2% mortality associated with palliative modified Blalock-Taussig-Thomas (BTT) shunt and 7.8% mortality for primary neonatal repair [1]. Thus, in many centers, alternative palliative options for promoting pulmonary arterial blood flow and growth have been evaluated, including stenting of the arterial duct [2], pulmonary balloon valvuloplasty [3], and, more recently, stenting of the right ventricular outflow tract (RVOT) [4-10]. Pulmonary balloon valvu-

loplasty has not gained widespread acceptance, as subvalvar muscular obstruction has been shown to be unresponsive to ballooning alone; however, in some substrates with predominant valvar obstruction, this option remains [11-13]. RVOT stenting was initially described by Gibbs et al. [4]; however, a larger, more contemporary series of 52 patients reported by Stumper et al. [5] demonstrated excellent outcomes. The authors concluded that RVOT stenting is a viable first-line treatment option for selected patients with severe RVOT obstruction as an alternative to systemic-to-pulmonary artery shunting procedures and for whom early complete repair carries significant additional risk or complexity. Only one patient in this cohort weighed < 2 kg. Despite significant advancements in stent design and profile to support stent delivery in low birth weight (LBW) infants, this cohort of patients remains a challenge, with a recent report suggesting a procedural mortality of 12% associated with interventions in infants < 2 kg [14].

In the past 4 years, we have placed 58 RVOT stents in 53 patients, with 4 patients weighing \leq 2 kg. Initially, we pursued a percutaneous approach in all patients; however, we have now moved to a periventricular approach through a small subxiphoid incision for patients < 2 kg. This approach provides a more direct route to the RVOT and possibly lessens the likelihood of hemodynamic instability induced by splinting the tricuspid valve. This report outlines these cases and



the learning curve associated with RVOT stenting in LBW infants.

Case Presentation

Case 1

An ex-36-week male infant weighing 1.9 kg was diagnosed soon after birth with ToF. There was failure to pass meconium, and a diagnosis of Hirschsprungs disease required a defunctioning colostomy. Due to progressive cyanosis, the infant was taken to the catheter laboratory in the second week of life and underwent RVOT stenting with a 4 × 12-mm coronary stent (Boston Scientific, Marlborough, MA, USA). There was severe RVOT narrowing (Figure 1) that was crossed with a 0.014" BMW wire (Abbot Vascular, Clonmel, Ireland) through a 4-F angled Terumo catheter (Terumo Europe NV, Leuven, Belgium) positioned in the RVOT. Further angiography was possible through a Tuohy-Borst placed on the catheter over the wire. Stent position was confirmed with the use of transthoracic echocardiography (TTE). The infant tolerated the procedure well with no hemodynamic instability following placement of the coronary wire in the distal right pulmonary artery. Oxygen saturation improved from mid-70% to mid-90%. The infant required further dilation of the stent at 3 months with a 6 × 20-mm balloon (Boston Scientific, Marlborough, MA, USA) and underwent uncomplicated complete surgical repair at 6 months.

Case 2

An ex-30 + 3-week male infant with a birth weight of 1.18 kg and an antenatal diagnosis of ToF became progressively cyanosed over the first 4 weeks of life. Despite administration of oxygen and propranolol for hypercyanotic spells, the patient remained symptomatic and was listed for urgent RVOT stenting at 4 weeks of age weighing 1.6 kg. The RVOT was crossed with a 0.014" BMW wire, and a 4 × 12-mm coronary stent was placed across the RVOT. Repeat angiography suggested residual infundibular muscle proximal to the RVOT stent. With advancement of a second stent, the initial stent milked distally into the main pulmonary artery. It was not possible to advance the second stent into a suitable position, and on removal through the sheath, the stent embolized off the

balloon into the right atrium. The stent was retrieved with an Amplatz goose neck snare (Covidien, Plymouth, MN, USA) and removed. Subsequently, a third 4 × 12-mm stent was advanced to the RVOT, covering the infundibular muscle, and was successful in alleviating the obstruction. The patient recovered well from the procedure without any sequelae.

At 8 weeks of age, weighing 2.2 kg, the patient re-presented with hypercyanotic spells, and echocardiography suggested muscle beneath the previously placed stents. Initial angiography confirmed the wedge of muscle. The stents were crossed with a 0.014" GrandSlam wire (Asahi Intecc, Osaka, Japan) and a 5 × 12-mm Formula 414 stent (Cook Medical, Bloomington, IN, USA) was placed in a good position. Final angiography and echocardiography suggested that the residual muscle bundle was fully covered by the stent.

Further subsequent desaturations led to a right modified BTT shunt at 10 weeks of age and subsequent complete repair at 26 weeks, weighing 5.3 kg.

Case 3

An ex-35 + 4 week male infant with a birth weight of 1.52 kg was diagnosed with ToF with severe RVOT obstruction following birth and was commenced on propranolol and prostaglandin. Clinical features were suggestive of Cornelia de Lange syndrome. At day 21 of life, weighing 1.84 kg, the patient developed progressive cyanosis despite prostaglandin therapy with a TTE, confirming progressive restriction of the ductus arteriosus. Following a multi-disciplinary team discussion, he was brought to the cardiac catheterization laboratory at 23 days of age for an urgent RVOT stent. The RVOT was crossed with a 0.018" Terumo wire through a 4-F non-tapered angled Terumo catheter. The Terumo wire was exchanged for a 0.014" GrandSlam wire, which led to significant hypotension thought to be secondary to splinting of the tricuspid valve. Some improvement was seen with the administration of adrenaline and withdrawal of the 4-F catheter. A 4.5 × 13-mm coronary stent was deployed under TTE guidance across the RVOT. This stent migrated proximally with balloon withdrawal, and attempts to manipulate it back into position with a balloon were unsuccessful. A further 4.5 × 18-mm coronary stent was advanced to stabilize the first stent; however,

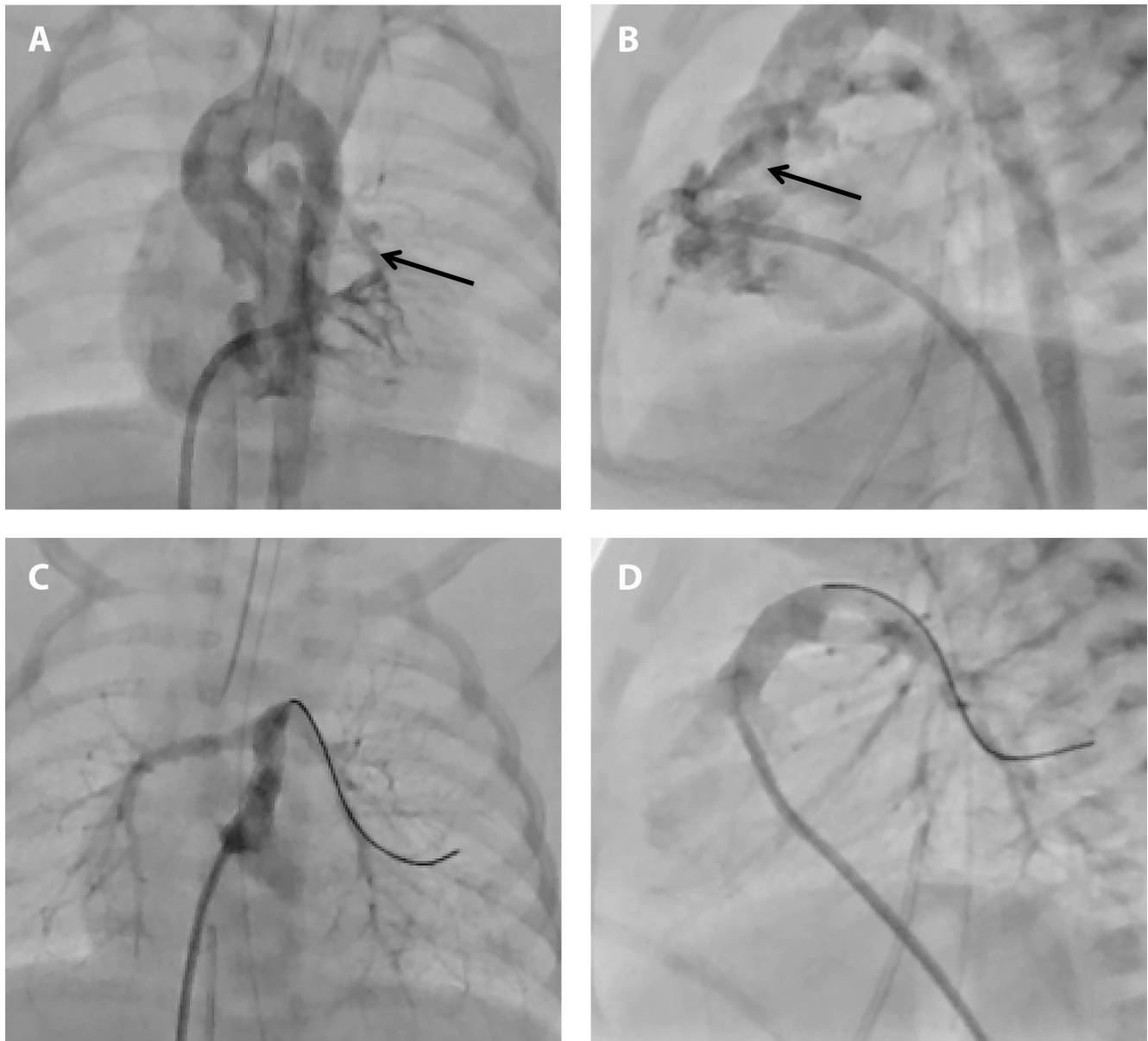


Figure 1. Panels A and B. Initial right ventricle angiogram demonstrating severely hypoplastic RVOT in both (Panel A) frontal and (Panel B) lateral planes (**black arrows**). Panels C and D. Repeat angiography following placement of a 4-mm coronary stent in the RVOT demonstrating (Panel C) improved filling of the pulmonary arteries and (Panel D) a more patent RVOT.

upon deployment it too migrated proximally into the right ventricle. The patient was stable initially, but further attempts to reposition the stents into the RVOT were unsuccessful and led to the patient becoming hypotensive and acidotic.

After the patient stabilized, a decision was made to perform a sternotomy and attempt to retrieve the stents through a 9-F sheath inserted into the right

ventricle. Through this, an Amplatz Goose Neck Snare was advanced into the right ventricle; however, attempts to snare the embolized stents led to damage to the right ventricular myocardium. The patient was emergently placed on cardio-pulmonary bypass (CPB) with retrieval of the stent and a modified systemic-to-pulmonary artery shunt performed. Despite this, the patient subsequently could not be weaned

from CPB due to severe biventricular dysfunction. He was felt to be too small for extracorporeal support and did not survive weaning of CPB.

Case 4

An ex-36 + 4-week infant with an antenatal diagnosis of ToF was born with a weight of 2 kg. He had the associated features of right undescended testes, hypospadias, micrognathia, butterfly vertebrae, and transient hyperinsulinemia.

The patient was listed for insertion of a RVOT stent on day 12 of life due to worsening oxygen saturation requiring re-commencement of prostaglandin to keep baseline saturation above 70%. On this occasion, initial right ventricle angiography was performed using a right femoral venous approach (Figure 2A and 2B). Similar to the third case, placement of a coronary wire across the tricuspid valve led to hypotension; therefore, on this occasion, a decision was made to revert to a subxiphoid approach to minimize hemodynamic instability. Through a small subxiphoid incision, a 5-F sheath was placed through the anterior surface of the right ventricle (Figure 2C). Following sheath angiography, a 0.014" BMW wire was placed in the distal right pulmonary artery through a 4-F Terumo catheter, and a 5 × 16-mm Formula 414 stent was deployed in a good position. Oxygen saturation increased to low 90%.

The patient had an uncomplicated post-operative course and recovered well. Ten weeks following the initial procedure, weighing 3.14 kg, he underwent uncomplicated implantation of a further 6 × 20-mm Formula 414 stent via the right femoral vein for progressive cyanosis secondary to muscle hypertrophy proximal to the RVOT stent. He subsequently underwent an uncomplicated full surgical repair at 5 months, weighing 5 kg.

Discussion

Despite significant advances in neonatal CPB, the mortality for symptomatic neonates with a diagnosis of ToF weighing < 2 kg has been reported as high as 49% [15]. This reflects the difficulties of CPB in this age group, with infants having smaller circulating blood volumes, higher oxygen consumption rates, and highly reactive pulmonary vascular beds. In ad-

dition, infants have labile thermoregulation and immature organ systems with multiple implications for ischemic tolerance and inflammatory response [16]. However, despite these difficulties, adopting a watch-and-grow approach has not been shown to improve outcomes [17, 18].

The unpredictability of pulmonary balloon valvuloplasty combined with the high mortality of both modified BTT shunt placement and primary repair signaled the need for an alternative palliation option—the RVOT stent. Since the initial report in 1997, numerous case series have reported on the use of RVOT stents in the palliation of ToF patients [4-10]. These mainly involved patients weighing > 2.5 kg.

In this series, we describe the evolution of our approach to insertion of an RVOT stent in ToF patients weighing ≤ 2 kg. In four patients, we performed RVOT stenting to facilitate somatic and pulmonary artery growth to an adequate size suitable for primary repair. Previous reports demonstrate that mortality associated with early primary repair is highest in neonates with small pulmonary arteries (i.e., Nakata index < 150 mm/m²) [19].

We have fine-tuned our approach to obviate the need for a long sheath to deliver the stent, which assists the approach in smaller infants; however, the potential hemodynamic instability seen with a stiff coronary wire across the tricuspid valve may limit even this approach. Following the relative ease of stent delivery in the initial case, we felt that that a coronary stent could be delivered without much difficulty. Lower profile and shorter length stents appeared to course through the smaller heart more easily. However, our second case highlighted the difficulties that can arise with percutaneous stent insertion, particularly ensuring adequate stent length without the availability of a long sheath, and the challenges with crossing a pre-existing stent in the RVOT. In the third case, hemodynamic instability required rapid stent deployment that ultimately led to stent migration. This case highlights the limited room for error and intolerance to stent manipulation within small hearts. In the fourth case, a hybrid perventricular approach via a subxiphoid incision was performed. This perventricular approach was previously described as a viable option in LBW neonates [10] and in this case allowed the infant to grow to successful neonatal repair.

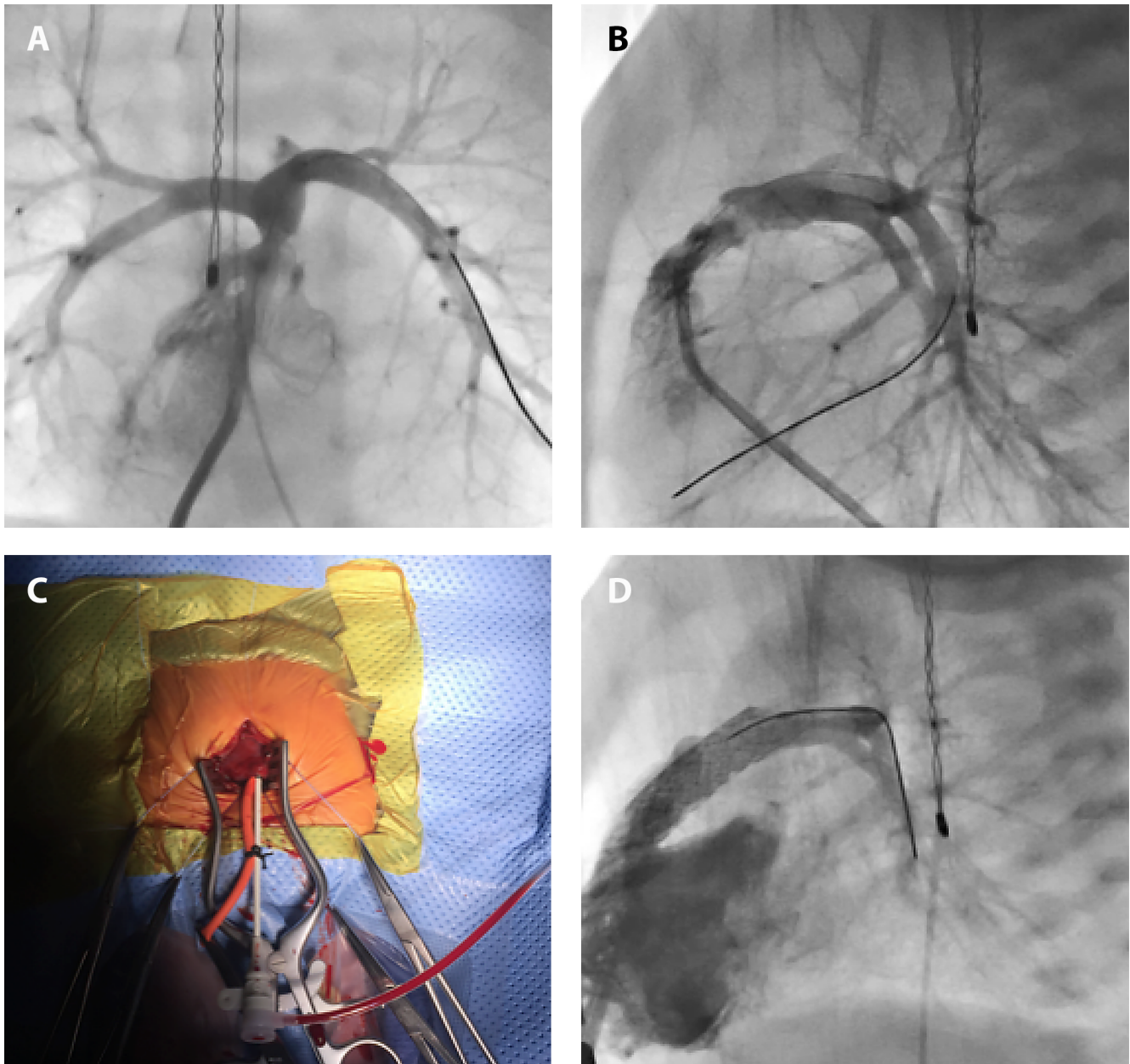


Figure 2. Panels A and B. Initial right ventricle angiogram demonstrating RVOT and pulmonary valve narrowing in both (Panel A) frontal and (Panel B) lateral planes. Panel C. 5-F short sheath paced through a small subxiphoid incision. Panel D. Right ventricle angiogram following stenting of the RVOT with a 5 × 16-mm stent.

Ultimately, these cases reflect a learning curve with RVOT stenting in infants weighing < 2 kg. Although an uncomplicated outcome is possible with a standard transcatheter approach, the margins of error are small, and complications are poorly tolerated. We believe a hybrid approach through a small subxiphoid incision provides the most direct route to the

RVOT in small infants and reduces hemodynamic instability, which may allow time to assess stent position adequately.

In conclusion, this case series highlights the evolution of an approach to RVOT stenting in infants weighing < 2 kg. Although a successful outcome with a transcatheter approach is possible, complications

may occur and are poorly tolerated. We feel a hybrid approach provides the most direct route to the RVOT with the least hemodynamic instability, providing concurrent angiography through the delivery sheath to facilitate optimal stent position. This small (n = 4) case series may not provide sufficient experience to support a periventricular approach as first-line for all infants ≤ 2 kg requiring RVOT stenting. However, early conversion to this approach should be considered in those infants who do not tolerate attempts at percutaneous stent delivery.

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Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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Cite this article as: Linnane N, Walsh KP, McGuinness J, Kenny D. Evolution of Approach to Right Ventricular Outflow Tract Stenting in Infants ≤ 2 Kgs. *Structural Heart Disease.* 2018;4(2):50-55. DOI: <https://doi.org/10.12945/j.jshd.2018.045.17>