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Stenting of the right ventricular outflow tract as primary palliation for Fallot-type lesions

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Abstract

Background: To describe the institutional experience, technical aspects and outcome of stenting of the right ventricular outflow tract (RVOT) in the initial palliation of symptomatic patients with severely limited pulmonary blood flow.

Methods: Retrospective case note and procedure review of patients undergoing stenting of the RVOT over a 10 year period at a quarternary institution.

Patients: Between 2005 and 2014, 76 selected patients underwent cardiac catheterization with the aim to implant a stent into an obstructed RVOT to improve pulmonary blood flow. Median age at stent implantation was 57 (range 4–406) days and median weight was 3.4 (1.7–12.2) kg.

Results: Seventy-two patients underwent stent implantation. Median procedure time was 53 (23–260) and fluoroscopy time 14 (5.2–73) minutes. Stents were implanted through either 4 F or 6 F sheaths. Median stent diameter was 5 (4–7) mm and stent length 16 (12–24) mm. There was one procedural death (1.4%) and one emergency surgery (1.4%). Saturations increased from 70 (52–83)% to 93(81–100)% [$p < 0.001$]. Within 30 days, two patients required early shunts due to inadequate palliation and two died from non-cardiac causes.

Conclusion: Stenting of the RVOT is an effective treatment option in the initial palliation of selected patients with very reduced pulmonary blood flow due to severe right ventricular outflow tract obstruction.

Keywords: Tetralogy of Fallot, Right ventricular outflow tract obstruction, Stent, Congenital heart disease

Background

The initial management of patients with normally related great arteries and a narrow right ventricular outflow tract (RVOT) with reduced pulmonary blood flow, as classically seen in Tetralogy of Fallot, remains challenging. True neonatal repair of these lesions remains the exception [1–3]. There is concern about increased early mortality and the universal need for transannular patching in this age group. The creation of a Blalock Taussig (BT) shunt is well established, but continues to have a high early and late complication rate and mortality [4].

Transcatheter techniques in the initial palliation of these patients have previously been attempted [5–8], but did not gain widespread acceptance. The medium term results after balloon valvuloplasty were largely unpredictable and the technical failure rate and complications with early attempts to stent the right ventricular outflow tract were not attractive.

It is only recently that several groups have revisited stenting the right ventricular outflow tract [9–12] in the initial palliation of symptomatic patients, including neonates.

This report summarizes institutional experience and lessons learned with stenting of the right ventricular outflow tract in an attempt to accelerate the learning curve of other centres considering this approach.

Methods

Between 2005 and 2014, seventy-six patients with normally related great arteries, a ventricular septal defect and a severely narrow right ventricular outflow tract resulting in profound desaturation were considered for stenting of the right ventricular outflow tract. Cases were selected after discussion within a multi-disciplinary team meeting.

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Initially cases were selected due to significant associated comorbidities or syndromes, then cases with very low bodyweight, in whom BT shunt would have carried very high risk, and then cases with severely hypoplastic pulmonary arteries or those with associated atrioventricular septal defect (AVSD). From 2011 onwards stenting of the RVOT has become the primary mode of palliation in symptomatic patients with tetralogy of Fallot morphology in our institution.

All procedures were carried out under general anaesthesia and mechanical ventilation. Emergency drugs prepared prior to the procedure were Propanolol 0.1 mg/kg in 10 mls, and Phenylepinephrine solutions 1 in 10,000. High inspired oxygen concentrations were used in all. Prostaglandin infusions (12 patients) were continued throughout the procedure.

Patients were positioned on the table with the arms elevated to allow for biplane angiography. A warming system (Bair Hugger, 3 M, Minnesota, USA) was used in all to prevent hypothermia. The chest and the upper abdomen of the child were covered with a sterile dressing (Opsite, Smith&Nephew, Florida, USA) to allow for intraprocedural echocardiographic evaluation using a standard ultrasound probe in a sterile sleeve. Right femoral venous access was the preferred approach in children weighing more than 2.5 kg. In smaller children a right internal jugular approach was frequently chosen. All patients received 50 iU Heparin/kg and standard antibiotic prophylaxis.

The initial right ventricular angiogram was performed through a diagnostic catheter placed towards the apex of the right ventricle under 30° RAO with 20° cranial tilt and a straight lateral projection (Toshiba Infinix CFi, Toshiba Medical Systems, Japan) (Fig. 1a). Following the angiogram the diagnostic catheter was withdrawn from the right ventricle and placed in the superior vena cava. Measurements of the length and diameter of the right ventricular outflow tract, pulmonary valve annulus and branch pulmonary arteries were made and compared to previous or simultaneous ultrasound measurements. Angiographic reference images were selected and displayed on the in-room monitors. If the decision to proceed to stenting the RVOT was made, all material was selected and prepared for use prior to entering the outflow tract or the branch pulmonary arteries.

Selection of the size and the type of stent to be implanted was guided by the size of the patient, the dimensions of the outflow tract and the anticipated length of palliation. Preference was to avoid crossing the pulmonary valve annulus with the stent. In small children in whom only short term (3–6 months) palliation was required, preference was to use a coronary stent (Liberte, Boston Scientific, Natick, MA, USA) (Fig. 1b). In larger patients, or in those with required medium to longer-term palliation, a bare metal peripheral vascular stent was selected. Initially, preference was for a JoStent (Abbot Vascular, Maidenhead UK). These are no longer in production. The Genesis stent (Cordis Corp, Miami Lakes FL) is an alternative, but suffers from significant shortening during placement. Of late, preference is for the Cook Formula pre-mounted 414 or 418 stent (Cook Europe, Bjaeverskov, Denmark), which does not shorten but does not easily conform to the curved anatomy of the outflow tract [13].

After selection of the stent to be used, the appropriate delivery sheath or guide catheter were selected. For coronary stents a 4 French (F) Flexor sheath (Cook Europe, Bjaeverskov, Denmark) or a 60 cm 6 F right Judkins guide catheter (Cordis Corp, Miami Lakes FL) were selected. Cook Formula stents were implanted through either 5 or 6 F Flexor sheaths. The delivery sheath was placed in the superior vena cava and a 4 or 5 F Right Judkins catheter was advanced past the tip of the delivery sheath. A rotating haemostatic valve was placed on the hub of the Judkins catheter, connected to the pressure line and side arm contrast syringe. A 0.014" coronary wire (Thruway, Boston Scientific, Natick, MA) was placed through the haemostatic valve near the tip of the

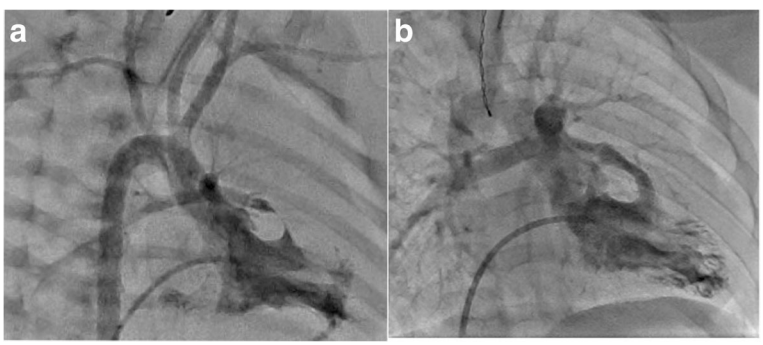

Fig. 1 **a**: Severe Tetralogy of Fallot in a 2.1 kg child with right aortic arch. The initial right ventricular angiogram is performed in 30 RAO and 20 cranial, demonstrating the severe anterior deviation of the outlet septum and the very small branch pulmonary arteries. **b**: Angiogram after stent implantation (4.5 mm × 16 mm coronary stent) sparing the pulmonary valve. There is good coverage of the proximal infundibulum and improved flow to the branch pulmonary arteries

catheter. Withdrawing the whole assembled system, the right ventricle was re-entered under pressure monitoring and the right ventricular outflow tract was intubated with the catheter. Side arm test injections were used to confirm the position of the catheter, rather than the wire. The catheter was then advanced to the distal branch pulmonary artery, under continuous pressure recording and repeat test angiogram. The coronary wire was placed in the distal branch pulmonary artery and the delivery sheath or guide catheter was advanced over the diagnostic catheter into the distal branch pulmonary artery. Then the diagnostic catheter was removed over the coronary wire and the sheath was aspirated and flushed. The pre-mounted stent was placed on the wire and advanced to the intended position within the RVOT. The stent was partially uncovered and repeat side arm test injections were performed. The stent was fully uncovered when the position was judged satisfactory [see attached Additional files 1, 2, 3, 4 and 5]. The delivery balloon was inflated manually with one hand whilst the other hand controlled the position of the stent system. Following placement of the stent, the balloon was slowly deflated whilst the delivery sheath was advanced over the balloon, so as to re-sheath it. This allowed for a secure position of the tip of the delivery system to be placed within the stent. Repeat test angiogram and brief ultrasound evaluation were carried out. In all cases the balloon was then advanced across the pulmonary valve for valvuloplasty. Oxygen saturations were monitored continuously.

Repeat cardiac ultrasound evaluation and right ventricular angiography was carried out (Fig. 2). A repeat blood gas analysis was obtained. Prostaglandin infusions were stopped. Finally the coronary wire was removed through the delivery sheath under fluoroscopic control, the delivery sheath was removed and manual haemostasis applied.

Patients were routinely extubated on the table and transferred back to the ward. Patients who experienced an increase of oxygen saturations in excess of 20% were commenced on twice daily diuretics. Aspirin medication (3–5 mg/kg) was commenced once the patient was feeding. This was maintained until complete repair with removal of the stent.

Results

Seventy-two patients underwent stenting of the RVOT. In 4 cases early on during the series the anatomy was judged to be unsuitable for stenting, due to a very short infundibulum (1 patient), presence of severe supravalve stenosis (2 patients) and/or improvement with balloon valvuloplasty on its own (2 patients). Age at stent implantation was a median of 57 (range 4–406) days. Median weight was 3.4 (range 1.7–12.2) kilograms.

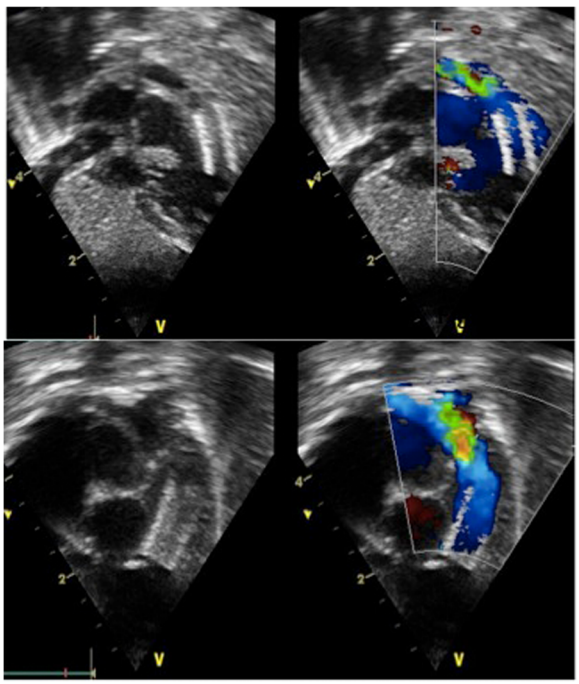

Fig. 2 Intraprocedural ultrasound study using subcostal (*top panel*) and parasternal views (*bottom panel*). Studies confirm that there us good coverage of the proximal infundibulum and no new tricuspid or aortic regurgitation. Note the guide wire is still in place (*bottom panel*)

Twenty-seven patients (37.5%) weighed less than 3 kg, with 15 children weighing less than 2.5 kg. There were 41 male patients.

Median procedure time was 53 (23–260) minutes and median screening time was 14 (5.2–73) minutes. Procedure time shortened significantly during the learning curve period. Median stent diameter was 5 (4–7)mm and median stent length was 16 (12–24) mm. Eight patients required two stents to cover the entire length of the infundibulum. Five of these were amongst the first ten patients treated with this new technique.

There was one procedural death (1.4%) due to perforation of the branch pulmonary artery with significant haematothorax and pericardial effusion. There was one emergency surgery (1.4%) for perforation of the right ventricular outflow tract. Two patients required early BT shunt for persistent desaturation despite the stent (2.8%). One patient required early complete repair for significant tricuspid valve regurgitation sustained during the stenting procedure. One patient required aortic valve repair after retrieval of the stent which had embolised to the descending aorta. Two patients died from non-cardiac causes.

Oxygen saturations increased from a median of 70(51–83)% to 93(81–100)% [$p < 0.001$].

Hospital stay, post procedure time of ventilation, PICU requirement (22% versus 100%, $p < 0.001$), post

procedure morbidity and mortality was significantly lower compared to a group of patients undergoing BT shunt at our institution.

There was significant growth of the branch pulmonary arteries over time. In a subgroup of patients with true tetralogy of Fallot the growth of the branch pulmonary arteries was found to be significantly better after RVOT stenting compared to BT shunts. The benefit of RVOT stenting was +0.59 z-score for the right pulmonary artery and +0.48 z-score for the left pulmonary artery.

The rate of catheter re-intervention after RVOT stenting was high (27/72 patients (37.5%). The majority of patients requiring redilatation of the stent or a second stent were those who were initially very low weight, and those who had associated lesions such as MAPCAs, hypoplastic pulmonary arteries or an AVSD. Re-intervention was performed in an attempt to increase the size of the stent and to provide longer term palliation by augmenting pulmonary arterial blood flow.

During subsequent complete repair in 51/72 patients (71%), the stent was only partially removed in the majority of cases. Mostly the back wall of the stent was left in situ. There was no mortality associated with complete repair.

The rate of transannular patching and the rate of conduit implantation did not differ significantly to the group of patients who had received a BT shunt as initial palliation in our institution. Neither were there significant differences in bypass or X-clamp time (data submitted for publication elsewhere).

Discussion

Creation of a BT shunt remains a very effective technique in the initial palliation of complex congenital heart disease. Nonetheless, the BT shunt continues to carry a very high early and late mortality and morbidity [4]. In addition there remains concern about potential for pulmonary artery distortion, the need for a separate surgical incision and long-term lung function when performed from a thoracotomy [14, 15]. Complete repair of tetralogy of Fallot in symptomatic neonates and young infants remains the exception in current clinical practice [2, 3].

The transcatheter management of congenital heart disease has dramatically evolved over the past two decades. Modern coronary interventional kit is suitable for catheter interventions in even premature neonates. As a consequence of this, and with the experience of stent implantation into the patent arterial duct or stenosed BT shunts [16, 17], there has been a re-consideration of stenting the right ventricular outflow tract in patients with tetralogy of Fallot after initial attempts by Hausdorff and Gibbs had been disappointing [7].

This paper summarizes the institutional experience and learning curve with stenting the RVOT in symptomatic patients with severe Fallot physiology. Patients were not randomized to this treatment, but rather were selected for a novel procedure on clinical grounds and on surgical risk assessment for the established BT shunt procedure. Over the past ten years this technique has evolved considerably and numerous technical modifications have been introduced from careful analysis of the problems and complications encountered. The technique has now become reproducible and of late has become our standard approach to patients with severe Fallot-type lesions that are deemed too young or too high risk for complete repair upon presentation. These constitute 17% of all Fallot patients in our practice. At the same time, surgical techniques have evolved to deal with the stented right ventricular outflow tract at the time of deferred complete repair [18].

A large number of centres are now considering the option of stenting the RVOT in preference to BT shunting, in particular in low weight neonates [19, 20] and hybrid procedures have been reported in premature neonates [21].

Important learning points from our institutional experience are numerous:

The use of long sheaths or guide catheters is highly advisable, due to the facility to perform repeat side arm test injections prior to stent positioning and the reduction in the risk of stent slippage or doing damage to the tricuspid valve/conduction system by covering the stiff coronary wire at all times.

All angiographic measurements underestimate the length of the right ventricular outflow tract. The stent chosen for implantation should be one size up from the measured length. Covering the proximal portion of the RVOT is crucial. Most often reliance on ultrasound measurements to select the stent length is preferable.

Stenting across the pulmonary valve annulus can be avoided in most patients who do not exhibit severe supravalve stenosis – yet balloon dilatation of the pulmonary valve (after stent placement) is important/mandatory. Sparing the pulmonary valve leaves the potential to avoid transannular patching at the time of complete repair. Also it should be the preferred technique, in very young neonates with small pulmonary arteries and elevated pulmonary vascular resistance.

In patients with an associated atrioventricular septal defect, particular care has to be taken to find a clean passage from the right ventricle to the pulmonary artery avoiding the chordal attachments of the superior bridging leaflet. The absence of new right atrioventricular valve regurgitation should be confirmed on cardiac ultrasound prior to placement of the stent.

In patients with Fallot-type double outlet right ventricle, the right ventricular outflow tract is classically positioned

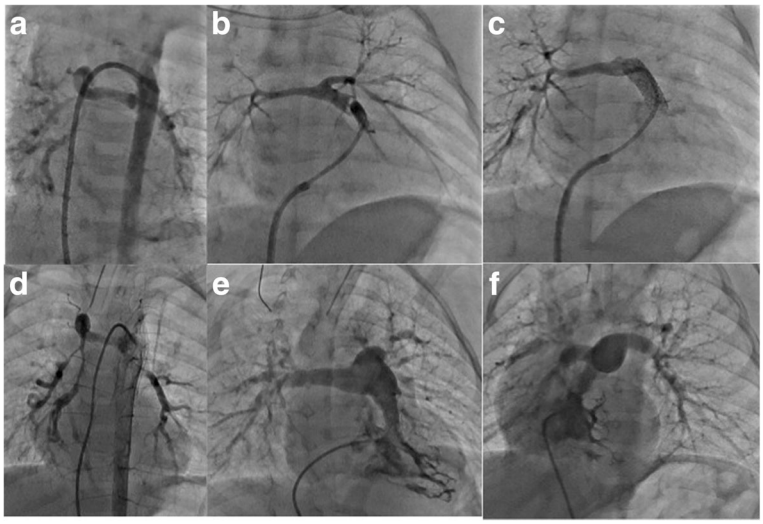

Fig. 3 Sequential angiograms in 2.3 kg child with severe Fallot, Alagille syndrome, hypoplastic pulmonary arteries and MAPCAs. **a**: Initial aortogram documents extensive MAPCAs with dual supply. **b**: RVOT angio documents severely hypoplastic branch pulmonary arteries and very dysplastic pulmonary valve. **c**: After stenting of the RVOT there is improved flow to the small pulmonary arteries. **d**: At follow-up angiogram 6 months later, there is documentation of very numerous long segment stenoses to the MAPCAs, and there has been very good growth of the central pulmonary arteries despite numerous peripheral stenoses (**e** and **f**). The child went on to have further dilatation of the stent and will eventually be listed for complete repair after embolization of MAPCAs

more horizontally. Care has to be employed to cover the entire length of the RVOT whilst selecting the most appropriate stent.

In patients with severe syndromes or permanent associated co-morbidities preference should be given to selecting a stent that can be significantly over-dilated without shortening at future re-intervention [13]. In some such cases stenting the RVOT may represent the best low-risk long-term palliation.

In patients with severe Tetralogy of Fallot and multiple aortopulmonary collaterals, the native pulmonary arterial system can be grown after stenting the outflow tract and then the collaterals can be taken care off by catheter embolization prior to complete repair (Fig. 3).

In developing countries with a high proportion of patients with severe Fallot physiology presenting late, stenting the RVOT may be a good means to reduce perioperative morbidity and mortality by gradually increasing pulmonary blood flow so as to reduce the risk of re-perfusion injury of the lungs during late one-stage repair.

Conclusion

Stenting the RVOT in patients with severe tetralogy of Fallot physiology has evolved to be a versatile and reproducible technique with superior outcome and complication rate compared to the BT shunt. It should be considered the first line palliation in patients who are not suitable or considered high risk for one stage complete repair and short term palliation for late presenters with markedly reduced pulmonary blood flow.

Additional files

Additional file 1: Movie 1. Initial right ventricular angiogram obtained in 30 degree right anterior oblique with 20 degree cranial angulation. (MOV 1590 kb)

Additional file 2: Movie 2. A 6 French right Judkins guide catheter is placed in the right atrium. A 4 French right Judkins catheter is used to cross the right ventricular outflow tract and to enter the right pulmonary artery using pressure monitoring and repeat test injections. (MOV 1316 kb)

Additional file 3: Movie 3. A coronary guidewire is placed in the distal right pulmonary artery, the Judkins guide catheter is advanced into the main pulmonary artery and the 4 French catheter is removed. The stent is delivered to the desired position and the Guide catheter is withdrawn to partially expose the stent. Further test injections are made. (MOV 980 kb)

Additional file 4: Movie 4. When the stent is placed in the desired position, it is fully uncovered and final test angiogram is performed before stent deployment. (MOV 1308 kb)

Additional file 5: Movie 5. After stent deployment, the pulmonary valve is dilated with the same balloon. A final angiogram and ultrasound is performed before removing the balloon and the wire. (MOV 1239 kb)

Abbreviations

AVSD: Atrioventricular septal defect; BT: Blalock taussig; F: French; iU: International units; MAPCA: Major aortopulmonary collateral arteries; PICU: Paediatric intensive care unit; RAO: Right anterior oblique; RVOT: Right ventricular outflow tract

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Availability of data and materials
Please contact author for data requests.

Authors' contributions
All authors read and approved the final manuscript.

Competing interests
The authors declare that they have no competing interests.

Consent for publication
See above.

Ethics approval and consent to participate
National and regional audit of stent use is compulsory in the United Kingdom. Consent for cardiac catheterization includes agreement to retain anonymised images and procedural data for later analysis and publication. No separate ethical approval was required for the study, as per regional guidelines.

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