

IMAGES

in PAEDIATRIC CARDIOLOGY

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MeSH

*Palliative Care; Pulmonary Valve Stenosis/*surgery; *Stents; Tetralogy of Fallot.

Traditionally, the management of infants with Fallot’s tetralogy (TOF) with excessively reduced pulmonary flow and cyanosis has been palliation until or unless complete repair is feasible. Palliation involves a procedure that augments pulmonary flow.

Most series recognise two subgroups of patients at high risk even in the current era: the cyanotic neonate/infant with small pulmonary arteries and those with complex anatomical variants of tetralogy and/or significant comorbidities such as additional congenital heart lesions (e.g associated Atrio Ventricular Septal Defect) or other congenital anomalies. There is an argument for palliation of these high-risk groups to allow for future potentially more effective elective repair.¹

Palliation in such cases has traditionally been by surgery, usually in the form of a surgical systemic to pulmonary shunt, such as a Blalock-Taussig (BT) or, less commonly, a limited right ventricular outflow (RVOT) patch. More recently, catheter interventions have played an alternative role by stenting the arterial duct (PDA) or the right ventricular outflow tract.^{2,3}

Ballooning of the right ventricular outflow tract is no longer practiced as the composite obstruction in TOF demands a device to support the dynamic obstruction.

Transcatheter stenting of the arterial duct carries relatively low risk but may actually result in significant morbidity and mortality including pulmonary artery deformation/stenosis and shunt obstruction.⁴ Moreover, it is less feasible to achieve controlled blood flow to the lungs and the PDA trajectory and shape can cause technical problems.

Transcatheter stenting of the RVOT⁵ is gaining popularity as this results in a more physiological haemodynamic result and encourages equal growth of small pulmonary arteries providing a better surgical substrate for subsequent repair.⁶

We present two Maltese babies who required early intervention using an RVOT stent and performed 10 years apart. Case 1 is a preterm baby (gestation 31/40) with TOF who required intervention due to increasing desaturation. Case 2 was performed in January 2005 and will be discussed later.

Case 1: One of twins with a birth weight of 1.25 kg born at 31 weeks gestation. She showed progressively deteriorating saturations despite propranolol at five weeks of age when she weighed 1.9 kg. The echocardiogram confirmed the diagnosis of situs solitus with Fallot’s tetralogy.

The pulmonary valve was very small and dysplastic but with a dynamic sub-pulmonary component. Catheterisation was carried out under GA entering the femoral vein percutaneously with a 4Fr Terumo Glidesheath. After haemodynamics and angiography (figures 1 and 2), the RVOT was measured at approximately 2-3mm by 10-11mm (figures 3-5). The RVOT was crossed with a Whisper 0.014” wire and a Judkins right guide catheter placed in the RVOT through which a Biotronik Pro-Kinetic 3.0x13mm stent was placed having pre-dilated the pulmonary valve due to resistance crossing this with the guide catheter or the stent. Deployment was done during apnoea to reduce movement but this also helps to ensure that all the team is engaged during this critical point of the procedure. The saturations improved immediately to the mid-90s % and there were no complications. Aspirin was commenced at 5 mg/Kg/day.

Figure 1: RV Angiogram in AP/Cranial 25 degrees showing Fallot’s tetralogy with composite RVOTO and a right aortic arch.



Figure 2: LV Angiogram in long axial view showing Fallot’s tetralogy with a single VSD and aortic override.



Figure 3: RVOT measurement

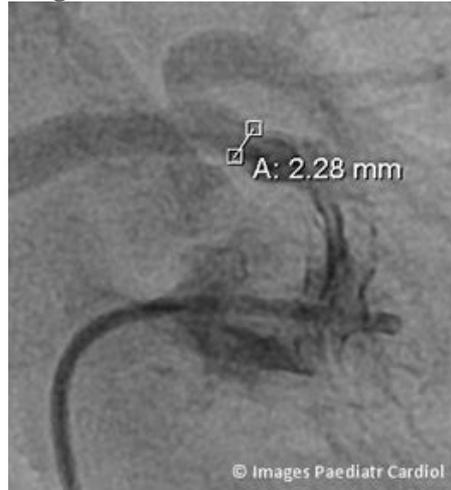


Figure 4: RVOT measurement

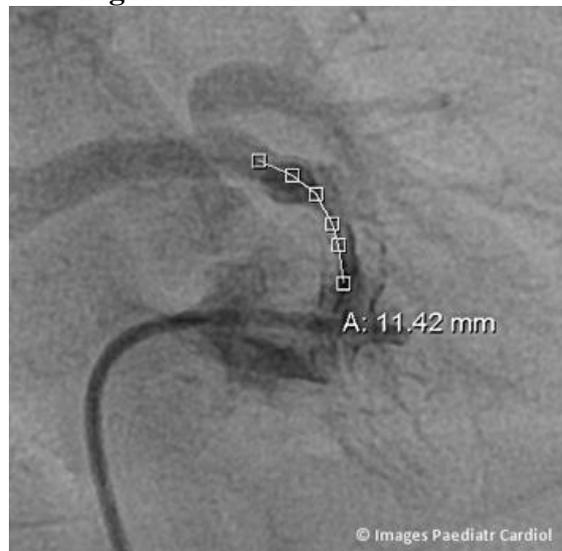


Figure 5: RVOT measurement



The stent was prepared (figure 6) inflated above nominal burst pressure giving an estimated diameter of 3.5mm due to the semi-compliant balloon (figures 7 and 8). As the pulmonary annulus was very hypoplastic and reparative surgery must entail a trans-annular patch, the stent covered the pulmonary valve as well as the infundibulum. When possible, the pulmonary valve should be preserved.

Figure 6: Stent prepared across RVOT

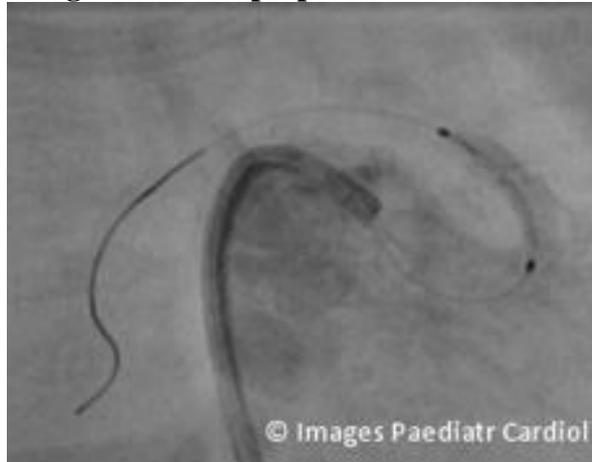


Figure 7: Stent inflated across RVOT



Figure 8: Post-procedure angiogram.



The echocardiographic appearance of the stent is shown in figures 9 and 10.

Figure 9: Echocardiographic appearance of stented RVOT in apical four chamber view.

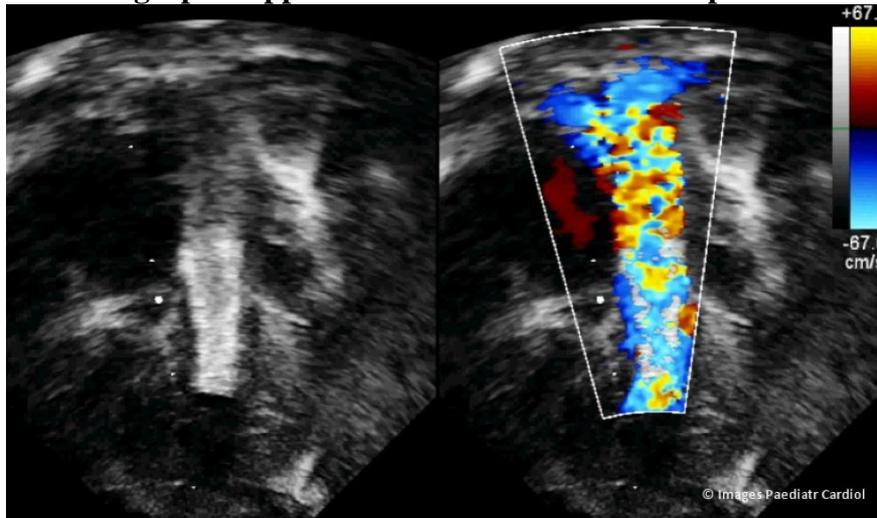
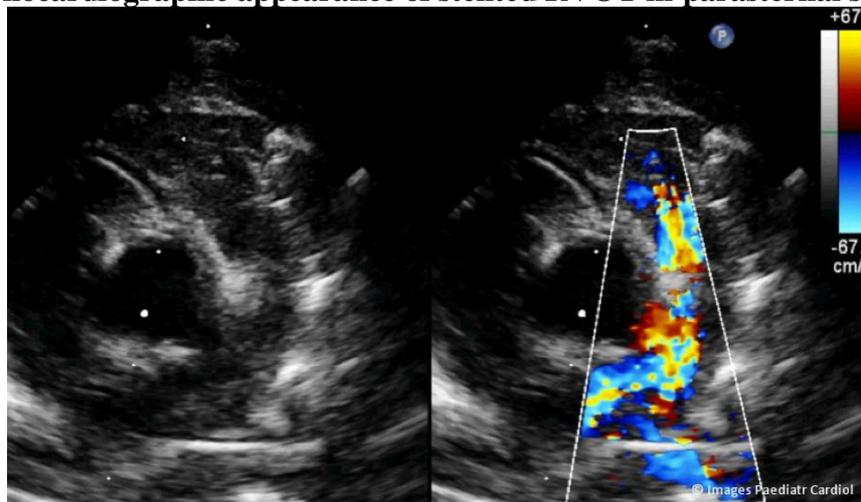


Figure 10: Echocardiographic appearance of stented RVOT in parasternal short axis view.



This stent will allow the baby to grow into a suitable weight for eventual Fallot surgery. Aspirin 5mg/kg/day was commenced, the baby was discharged home with a saturation of 80% off propranolol. If the patient develops cyanosis again during the follow up clinic he will be referred for reparative surgery if the weight and anatomy are suitable otherwise he may require dilatation of the current stent with the possibility of the addition of a second stent if the muscular obstruction extends below the bottom of the stent.

Case 2: This was a male baby who was born in November 2004 at 36 weeks of gestation, small for gestational age. He had Fallot’s tetralogy, a left aortic arch, left superior vena cava to coronary sinus and additional problems. These included omphalocele (see CXR, figure 11), a left inguinal hernia and talipes equinovarus.

Figure 11: Chest and abdominal x-ray showing large omphalocele



Karyotype was normal. He was catheterised at seven weeks of age due to increasing cyanosis; the latter was in part related to small lungs which became compressed after repair of the omphacocele but there was progressive worsening of the RVOTO. Weight was 3kg. Access was extremely technically difficult. Standard percutaneous approach on both groins failed and a cut down to the right femoral vein showed that this was fibrosed. Similarly, right subclavian vein and right internal jugular approaches failed. The superior vena cava was entered directly and a stent (Biodivisio 4x15 mm) across the RVOT decreased the gradient (peaks) from 90 to 55mmHg (figures 12-14). A Hickman line was left in place at the end of the procedure.

Figure 12: Pre-stent angiogram. Arrow indicates site of RVOTO.

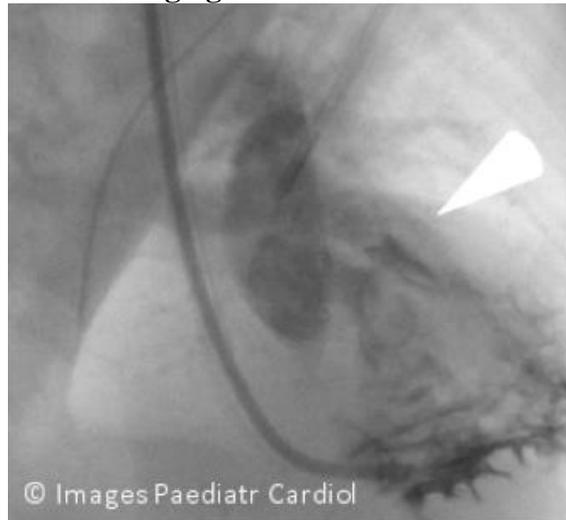


Figure 13: Stent inflation across the RVOT.

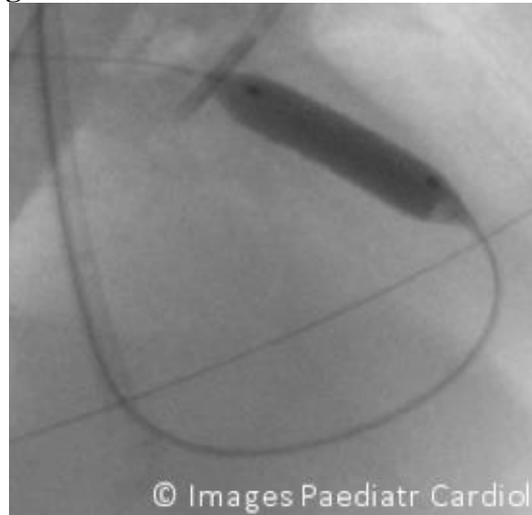
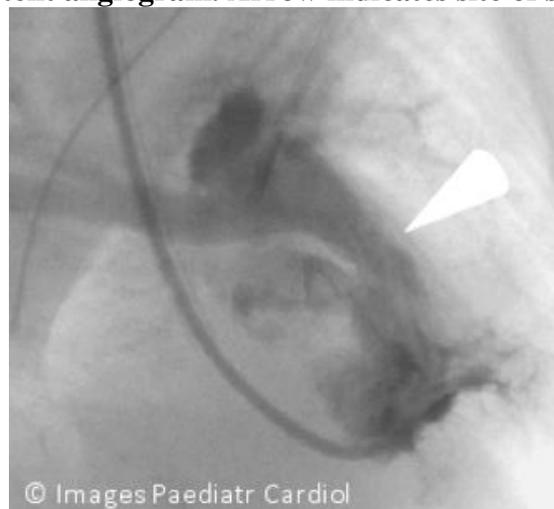


Figure 14: Post-stent angiogram. Arrow indicates site of stent implantation.



The baby remained initially well but had episodes of repeated desaturation requiring ventilation, despite a patent stent. There were concerns with regard to ongoing sepsis, chronic lung disease with

poor lung function and this was explained to the family. An ultrasound of the brain one month later showed cortical atrophy with an interhemispheric fissure >1cm wide, a periventricular cyst of the right hemisphere and biventricular dilatation, almost certainly due to hypoxic ischaemic encephalopathy. After discussion with the parents, support was withdrawn and the patient passed away shortly after.

In conclusion, RVOT stents provide a viable and logical alternative that is more haemodynamically physiological than traditional techniques, such as, shunts and arterial duct stenting, which are a haemodynamically equivalent procedure.

Potential problems with RVOT stenting include RVOT rupture, stent embolisation to pulmonary artery or aorta, stent fracture/compression and in-growth stenosis. However, stenting the RVOT as palliation leaves a virgin field for the surgeon during repair and does not have the failure rate of a BT shunt which is around 8%. The eventual repair takes slightly longer but with no clinical implications. Whereas in 2005, RVOT stenting was carried out in desperate situations,^{7,8} these days several centres use it as their palliation of choice for infants with TOF who are not suitable for repair at the time of presentation.⁶ In very small infants, a jugular approach may be preferable as this vein is larger than the femoral.

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