Stenting of right ventricular outflow tract in Tetralogy of Fallot with subarterial ventricular septal defect: A word of caution

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ABSTRACT

We report a case of Tetralogy of Fallot with severe cyanosis who underwent a successful right ventricular outflow tract stenting. Follow-up echocardiography revealed moderate aortic regurgitation due to the impingement of the stent on the aortic valve. The patient underwent successful surgical correction at which time the stent was removed completely with a resolution of the aortic regurgitation.

Keywords: Aortic regurgitation, ductal stenting, Tetralogy of Fallot

INTRODUCTION

Right ventricular outflow tract (RVOT) stenting has become an accepted initial palliative procedure for Tetralogy of Fallot (TOF).[1] Although primary repair in the neonatal period has been established[2] this remains a high-risk surgery particularly in low-weight neonates or those associated lesions such as atrioventricular canal defect, hypoplastic pulmonary arteries (PAs), coexisting morbidities or syndromes.[3] Although RVOT stenting has been shown to be safe,[4] various real and theoretical complications have been discussed with regard to use of stents in this context. However, there has been no report of stent protrusion impinging on the aortic valve causing aortic regurgitation. We describe a case wherein an RVOT stent caused aortic regurgitation in a patient with subarterial type of ventricular septal defect (VSD).

CASE REPORT

A 3-year-old female patient from a neighboring country with DiGeorge syndrome and TOF was referred to us with severe cyanosis and decreased effort tolerance. She was admitted with severe cyanosis with oxygen saturation of 70%. Echocardiography showed TOF anatomy, severe infundibular stenosis with a pressure gradient of 87 mmHg, a large subarterial type of VSD, and major aortopulmonary collateral arteries (MAPCAs) were seen. There was a trivial aortic regurgitation. Computerized tomography of thorax was carried out to delineate PA anatomy. This showed TOF with pulmonary stenosis, confluent PAs with a 3 mm right PA, a large collateral from the mid-descending aorta supplying the right PA and small collaterals from a left innominate artery supplying the left PA. Cardiac catheterization showed TOF anatomy with RVOT infundibular and valvular stenosis with the narrowest diameter at the infundibulum of 3.1 mm, small but confluent PAs, a small pulmonary annulus (3.6 mm) with thick cusps and 3 large MAPCAs. Stenting was done by first opening up the RVOT with a PowerFlex balloon 5 mm × 20 mm then the Express Vascular SD 7 mm × 19 mm stent was positioned across the RVOT. Poststenting angiogram showed good position of the stent and congested lung fields, which improved with medical management. Oxygen saturation improved to 88% and echocardiography showed a decrease of RVOT pressure gradient to 51 mmHg as well as good branch PA flow. There was mild aortic regurgitation noted.

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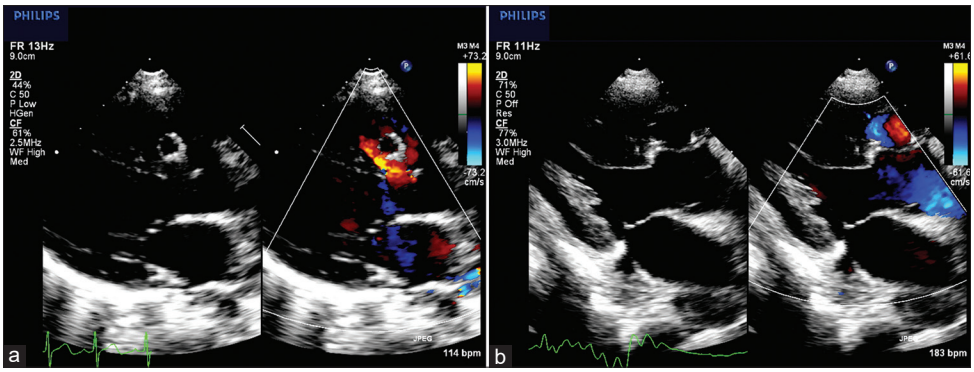

Figure 1: (a) Preoperative echocardiogram in parasternal long axis view shows stent en face view in close proximity to right coronary cusp. Color flow shows mild aortic regurgitation. (b) Postoperative echocardiogram in parasternal long axis view showing the resolution of aortic regurgitation

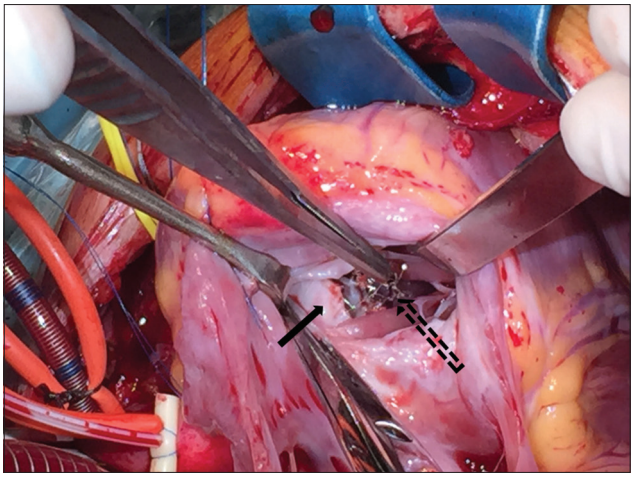

Figure 2: Intraoperative image of stent impinging on the aortic valve. Right coronary cusp with the impinged stent (bold arrow). Stent peeled from the aortic valve (dotted arrow)

The patient was admitted at age 4 for catheterization main PA size had increased to 7.2 mm from 6.7 mm, and branch PA size had also increased to approximately 5 mm. At 1 year follow-up, oxygen saturation was 87%. The gradient across the stent was 81 mm Hg. The prominent MAPCA was unable to be occluded due to its large size and hence decided to ligate it during the surgery. Echocardiography revealed aortic regurgitation of mild to moderate grade due to restricted movement of the right coronary cusp [Figure 1]. The child weighed about 10.2 kg at the time of stent placement and increased in weight to 13.8 kg before surgery.

Total correction of TOF with, monocusp reconstruction of the pulmonary valve, removal of the RVOT stent and MAPCA ligation was carried out on the same admission. The intraoperative findings were that of absent infundibulum with a fibrous continuity between the hypoplastic pulmonary annulus with the stent *in situ* and the aortic valve. This resulted in the stent impinging onto the right coronary cusp of the aortic valve restricting its movement [Figure 2]. The stent was removed with gently peeling it from the valve and leaving a small remnant on posterior wall of the pulmonary valve.

Postoperative echocardiogram showed good ventricular function with no residual VSD, a pressure gradient across the RVOT of 20 mmHg, and good flow into the left and right PAs. The aortic regurgitation was trivial [Figure 3]. Her postoperative course was uneventful. Follow-up is planned for this patient 1-year postoperatively showed no evidence of aortic regurgitation.

DISCUSSION

This case clearly illustrates the problems related to RVOT stent placement especially in patients with subarterial VSD. The incidence of subarterial VSD in TOF is 6%–8%.[5] This particular subset of TOF is more frequently seen in the Asian population where it has been noted to be around 10%–20%.[6] In this group of patients, the septomarginal trabeculae are situated anteriorly and inferiorly, and the ventriculo-infundibular fold is posteriorly forming the border of the VSD. The semilunar valves form the ceiling of the defect. The infundibular septum in this heart is small and mostly replaced by a bridge of fibrous tissue that separated the defect from actual pulmonary valve annulus. Furthermore, the pulmonary valve ring and main PA are also usually hypoplastic.[7] Due to this anatomy of deficient infundibulum with hypoplastic pulmonary annulus, the stent when placed in the RVOT had impinged on the native aortic valve restricting the movement of the valve leading to aortic regurgitation.

The reported complication rate of RVOT stenting is about 7%.[1] Furthermore, no stent-related complications such as ventricular arrhythmias, atrioventricular block, aortic regurgitation, myocardial ischemia, or stent fractures were reported from the previous series. One case of stent dislodgement with tricuspid regurgitation has been described in a case series.[8] Most of the patients with RVOT stent will require a transannular patch, and about half of the patients, the stent can be removed

completely.[9] A small part of the stent may be left *in situ* to avoid damage to the surrounding tissue and the margin of the VSD. In this case, the stent adhered to aortic valve and was removed leaving a small part on the posterior wall of the pulmonary annulus.

Although RVOT stent has become an established palliative procedure in TOF, one needs to be wary of the anatomical variations that may preclude its application. In the presence of subarterial VSD, which is associated with hypoplastic pulmonary valve, one will invariably perform a transannular patch for reconstruction at the time of total correction.[10] Hence, it is better to place the stent at the level of the annulus into the main PA. This will prevent the lower positioning of the stent, which could likely damage the aortic valve. Furthermore, in this setting, it is better to proceed to corrective surgery after a short period of palliation to prevent damage of the stent to the adjacent structures.

CONCLUSION

Although RVOT stenting is an accepted palliation for patients with TOF, consideration should be given to the anatomy for a safe placement of the stent especially when there is a deficient infundibular septum as in a doubly committed subarterial VSD.

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Conflicts of interest

There are no conflicts of interest.

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