Aortopulmonary Fistula After Outflow Tract Stent in Repaired Truncus

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We present a case of an iatrogenic aortopulmonary (AP) fistula in a 9-year-old patient with a history of repaired truncus arteriosus without the use of a right ventricle to pulmonary artery conduit and subsequent transcatheter placement of a right ventricular outflow tract (RVOT) stent. Redilation of the stent resulted in a defect in the aortic wall and the creation of an AP fistula with an associated hemodynamically significant left to right shunt. This case demonstrates a previously unreported adverse event of transcatheter RVOT reintervention after truncus arteriosus repair.


Persistent truncus arteriosus is a congenital heart defect caused by failure of the pulmonary artery trunk and aorta to separate, resulting in both great vessels coming off a common trunk with only one semilunar valve (truncal valve). Typically, repair consists of closure of the ventricular septal defect (VSD), incorporation of the truncal valve into the left ventricular outflow, detachment of the pulmonary arteries (PAs) from the common trunk, and creation of continuity between the right ventricle (RV) and the PA confluence. Various techniques have been proposed to reconstruct the RVOT. However, the vast majority of repairs involve placement of an RV to PA conduit, which is typically a homograft in the neonatal period. Although, conduits often work well for many years, they have a limited lifespan because of the development of stenosis, regurgitation, or both and need to be upsized as the patients grow. Barbero-Marcial and associates [1] proposed a conduit-free technique for establishing RV to PA continuity. In this technique, the PA confluence is not detached from the truncus, and a portion of the reconstructed RVOT is made up of the aortic wall. Patch material is used to complete the circumference of this reconstructed RVOT. It is important to note that when this approach is used, patients do not have two distinct outflow tracts [1].

Our patient was a 9-year-old boy with DiGeorge syndrome and truncus arteriosus (type A2 [2]) who had undergone surgical repair in the neonatal period without the use of a conduit according to the technique previously described by Barbero-Marcial and associates [1]. Upon transfer to our institution at 3 years of age, the patient had experienced evidence of RVOT obstruction, which was alleviated by transcatheter RVOT stent placement (19 mm Genesis; Cordis Corporation, Bridgewater, NJ). With the exception of slow weight gain, our patient remained asymptomatic, with normal activity levels. By age 9 years, recurrent severe RVOT obstruction was suggested by echocardiography. Therefore, he was returned to the catheterization laboratory, where his RV pressure was found to be moderately elevated (58 mm Hg with simultaneous systemic systolic pressure of 90 mm Hg). To reduce his RV hypertension, the stent was dilated with high-pressure balloons (14-mm and 16-mm balloons to nominal pressure (Bard Peripheral Vascular, Tempe, AZ). The patient tolerated this well, the stent diameter increased, and the RV systolic pressure decreased to 36 mm Hg. The procedure appeared to have been acutely successful, and the patient was discharged home the next day after an uncomplicated recovery. However, 2 months later he was readmitted because of a new onset of signs and symptoms consistent with biventricular failure, confirmed by new moderate depression of biventricular systolic function on echocardiography (previously normal). Cardiac catheterization was performed. Oximetry demonstrated a left to right shunt in excess of 2:1. The right and left ventricular end-diastolic pressures, which had been normal 2 months previously, were now severely elevated (22 mm Hg). Angiography demonstrated a new communication between the ascending aorta and the RVOT at the level of the previously placed stent, ie, an aortopulmonary (AP) fistula (Fig 1). Truncal valve function and coronary artery flow remained normal.

Transcatheter closure of the defect was considered, but a surgical approach was preferred. A redo sternotomy was performed after establishment of cardiopulmonary bypass through the femoral vessels. After adequate cooling, the circulation was arrested. The aorta was divided, and cardioplegia was administered. The fistula was easily identified just above the anterior leaflet of the truncal valve (Fig 2). The RVOT was opened, and the stent was removed. The defect in the aortic wall was closed primarily with pledgeted sutures of 5-0 Prolene and then oversewn with a second layer of 5-0 Prolene. The aorta was reanastomosed, and cardiopulmonary bypass was reinitiated. A 21-mm Perimount valve (Edwards Lifesciences, Irvine, CA) was implanted in the RVOT in a heterotopic position, and a 20-mm Hemashield graft (MAQUET, Wayne, NJ) was used to reestablish RV to PA continuity. The patient was separated from cardiopulmonary bypass, on low-dose epinephrine and dopamine, with satisfactory hemodynamics.

At follow-up, 5 months postoperatively, the patient was in New York Heart Association functional class II. Echocardiography showed moderate LV systolic dysfunction.

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LV ejection fraction 40%) compared with severe (LV ejection fraction 25%) in the early postoperative period.

Comment

This unusual case illustrates that the application of specific surgical techniques may predispose to unexpected adverse events after endovascular interventions. Common endovascular interventions after truncus arteriosus repair include angioplasty, stent placement, or both in stenotic RV-PA conduits to temporize surgical conduit revision. Given that the vast majority of truncus arteriosus repairs include RV-PA conduit placement, interventional cardiologists are very familiar with this procedure, being aware of the potential adverse events, including conduit fracture or even rupture (particularly if the conduit is heavily calcified).

Prior case reports of iatrogenic AP fistula exist in the literature, several of which have occurred after redilation of PA stents in patients after arterial switch operations for D-transposition of the great arteries [3-5]. These were managed by transcatheter interventions (e.g., endovascular stent graft or covered stent), the latter of which was considered in our patient but several factors led us to favor surgical repair. Our case highlights an unusual adverse event after reintervention on a previously placed RVOT stent in the context of repaired truncus arteriosus by use of the method described by Barbero-Marcial and colleagues [1], which is a conduit-free approach that does not create two distinct outflow tracts. The aortic wall constitutes part of the RVOT. Had the patient undergone a typical conduit repair, a scarred-down conduit wall would have likely provided structural integrity and support for placement and reexpansion of the stent. Inasmuch as this is an isolated case report, we cannot speculate about the risk of this adverse event. It should be noted that we have successfully treated several patients with similar anatomy using bare metal stents. However, we recommend a cautious approach to transcatheter treatment of obstruction in patients with this type of nonconduit RVOT reconstruction such as that described by Barbero-Marcial and colleagues or as used in the classic Nikaidoh operation. Careful graded angioplasty before stent placement, consideration of primary stenting with covered devices, and aortic root angiography perhaps at several phases during the intervention might aid in the early identification and perhaps prevention of this adverse event. Finally, after such RVOT interventions, a new AP fistula should be considered in the evaluation of new-onset heart failure in this patient group. Furthermore, our case highlights the importance of communication between surgeons and interventional cardiologists to ensure full understanding of potential adverse events related to applications of atypical surgical techniques.

In conclusion, we report the first iatrogenic AP fistula after transcatheter angioplasty of a previously placed RVOT stent in a patient with a nonconduit type repair of truncus arteriosus. We favor caution in the implantation of non-covered stents within the RVOT of such patients because we suspect that it may predispose to the development of
AP fistulae and subsequent clinical heart failure with potential irreversible ventricular dilation and dysfunction.

References


