Transcatheter management of aorto-right ventricular tunnel: A surprise in the catheterization laboratory

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ABSTRACT

Herein, we report successful device closure of aorto-right ventricular tunnel (ARVT) in a 2-year-old boy presenting with recurrent respiratory tract infection and poor weight gain. He was initially diagnosed with coronary arteriovenous fistula after the clinical examination and echocardiogram. However, his cardiac catheterization revealed ARVT. Although most of the aorto-ventricular tunnels are managed by surgical intervention, we successfully managed our case with transcatheter intervention.

Keywords: Aorto-left ventricular tunnel, aorto-right ventricular tunnel, arteriovenous fistula, device closure

INTRODUCTION

Aortic-ventricular tunnel is a rare congenital anomaly[1] with an extracardiac fistulous communication between the ascending aorta (AA), above the sinotubular junction (STJ), and one of the ventricles. The exact incidence is unknown, it ranges from 0.1% to 0.5% of congenitally malformed hearts.[2] The etiology of aorto-ventricular tunnel is uncertain. It appears to result from a combination of maldevelopment of the cushions, which give rise to the pulmonary and aortic roots, and abnormal separation of these structures.[2] Most of them develop symptoms of heart failure during the 1st year of life due to aortic regurgitation.[3] The onset, severity, and progression of heart failure vary and ranges from in utero fetal death[4] to asymptomatic adulthood.[5] Early diagnosis and surgical correction are essential to prevent irreversible myocardial dysfunction and heart failure. We report our experience of transcatheter management of a case of aorto-right ventricular tunnel (ARVT).

CASE REPORT

A 2-year-old boy was referred to our center with a history of recurrent respiratory tract infection since 4 months of age. On presentation, the child’s weight and height were 9 kg and 89 cm, respectively, with 100% arterial saturation in all four limbs. His cardiovascular examination showed a continuous murmur in the left lower sternal border. Normal heart size and increased pulmonary vascularity were noted on his chest X-ray [Figure 1]. His electrocardiogram (ECG) showed sinus tachycardia and an incomplete right bundle branch block [Figure 2]. His transthoracic echocardiogram (Echo) showed a large coronary arteriovenous fistula (CAVF) of 7.3 mm in diameter arising from proximal segment of the right coronary artery (RCA) and draining into the right ventricular outflow tract (RVOT) [Figure 3]. The child was taken up for cardiac catheterization to confirm the diagnosis and plan the further management. Since most of the CAVF are amenable to transcatheter closure (TCC), we preferred cardiac catheterization angiogram to computed tomography angiogram to avoid additional radiation. However, to our surprise, aortic root and selective coronary artery angiograms showed fistulous communication from the aorta, above the STJ to RVOT [Figure 4] with normal coronary artery origin and its course [Figure 5]. Hemodynamic data showed...
mildly elevated right ventricular and pulmonary artery systolic pressures (42 mmHg), significant left to right shunt (Qp/Qs – 1.5), and normal pulmonary vascular resistance/systemic vascular resistance ratio (0.3). With this cardiac catheterization data, we changed our diagnosis as a case of AVRT with a significant left to right shunt. Since the tunnel was well away from the coronary artery origin and aortic valve, we decided to perform a TCC instead of surgical correction. The tunnel was conical shaped with a length of 15 mm, and width of 10 and 3.6 mm at aortic and RV ends, respectively, with the RV end being the narrowest segment of the tunnel.

After discussion with family members, written consent was obtained for the procedure. Our plan was to use ADO II device of 1.5–2 mm more than the narrowest segment of the tunnel. Hence, we decided to use 6/4 mm ADO II device. After inserting 5F terumo sheath (Terumo corporation, Tokyo) in the right femoral artery, the tunnel was crossed using 260 cm 0.035” J tipped Terumo wire. A 6/4 mm ADO II device (AGA Medical Corporation, Plymouth, MN) was deployed in the tunnel through retrograde technique using 5F Judkins right guiding catheter. Check angiogram showed significant residual shunt despite repeated repositioning of the device [Figure 6]. In view of a significant residual shunt, ADO II device was retrieved, and an 8 mm muscular VSD device (through 6F Amplatzer sheath) was deployed. However, significant residual shunt persisted. Suspecting additional defects, we decided to cross the tunnel after occluding one defect with a device. So we redeployed the 6/4 mm ADO II device in one of the defects and then we crossed the second defect using 260 cm 0.035” J tipped Terumo wire from left femoral artery side. Using 5F Judkins Right guiding catheter, 5/4 mm ADO II device was deployed in the second defect. Check angiogram showed no residual shunt across the tunnel [Figures 7 and 8]. Postprocedure angiogram and Echo did not show coronary artery obstruction or aortic valve function interference by the devices. We waited for 30 min before releasing the devices. ECG was monitored throughout the procedure. Transthoracic Echo was used in addition to fluoroscopy as a guide to position the device during deployment. Echo assessment of device position, aortic valve function, and myocardial contractility was done before the release of both the devices. The total duration of the procedure was 3 h with a fluoroscopy time of 50 min. Predischarge Echo and ECG showed no residual shunt and no myocardial ischemic changes.
Aorto-ventricular tunnel is an extracardiac channel that connects the AA to the LV or RV, passing outside the heart into the tissue plane between the muscular subpulmonary infundibulum and the arteriovenous (AV) sinuses. As per the statistics available 90% of the aorto-ventricular tunnels are aorto-left ventricular tunnel and the rest are ARVT. In our literature review, 31 cases of ARVT were found, 2 cases underwent TCC (details of the TCC were not mentioned), and the remaining cases underwent surgery. It must be differentiated from a ruptured sinus of Valsalva, which is an intracardiac channel that connects a sinus of the AV to one of the cardiac chambers. It also differs from a coronary artery AV fistula, which is an abnormal connection between one of the coronary arteries to one of the cardiac chambers.

In our case, precatheterization diagnosis was coronary AV fistula because of the location of the shunt and the aneurysmal aortic end of the tunnel which obscured the RCA origin on Echo. A retrospective review of our preprocedure Echo did not give any clues to diagnose the ARVT. Although the number of TCC of AVT are less as compared to surgical repair, TCC is an effective alternative and a safe procedure in selected cases. One more interesting finding noted in our case is the unusual occurrence of two separate tunnels communicated from aorta to RV, and two devices were used to close the defects.

**CONCLUSION**

AVRT is a rare congenital heart defect, and it is treated conventionally by surgical repair. However, our case is an example of surprise on cardiac catheterization laboratory, and TCC of AVRT is possible and safe.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.
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