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CASE REPORT

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Complete percutaneous repair of Tetralogy of Fallot in adult: a case report

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Abstract

Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease and is characterized by an antero-superior deviation of the infundibular septum with a consequent large malaligned ventricular septal defect (VSD) and a pulmonary and sub-pulmonary (infundibular) stenosis. Surgical repair has been the cornerstone of treatment that is electively performed early in their lives between 3 and 6 months of age. With advancements in transcatheter interventions, the complete percutaneous repair of TOF, a complex disease with multiple treatable lesions, is becoming a conceivable possibility. Here, we report the case of total transcatheter correction of an 18-year-old boy with TOF, performed in two stages. The first stage involved addressing the right ventricular outflow tract (RVOT) obstruction with balloon pulmonary valvuloplasty (BPV) and occluding the conal artery using absolute alcohol and a coil. In the second stage, the VSD was closed with a Multifunctional Occluder (MFO) Konar device 14–12 mm (Lifetech, China). While surgical treatment remains the gold standard for total correction of TOF, the transcatheter approach can also be considered for selected group of patients who are surgically turned-down.

Case report

TOF consists of RVOT obstruction, malaligned subaortic VSD, overriding of the aorta, and right ventricular hypertrophy [1]. Total correction conventionally is performed by open heart surgery, where both the stenotic right ventricular outflow and VSD are repaired. The clinical spectrum of TOF varies from severe cyanosis and ductal-dependent pulmonary circulation to older patients with subtle cyanosis. Few cases of transcatheter complete repair have been reported so far [2, 3]. In the developed world, most TOF patients have a total surgical correction in early infancy, but in developing countries, unrepaired cyanotic heart defects including TOF continue to exist.

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We report a case of an 18-year-old boy who presented to our institution with subtle cyanosis and progressively worsening exertional shortness of breath. Physical examination and ECG were consistent with TOF. Chest radiography shows boot shaped heart and right-sided aortic arch. Echocardiography revealed 30% aortic overriding with a non-restrictive subaortic VSD of size 11 mm (Fig. 1A), with RVOT obstruction in the form of pulmonary valvular and infundibular stenosis with estimated pressure gradient of 110 mm Hg (Fig. 1B). Pulmonary annulus was 22 mm.

Cardiac catheterization and angiography confirmed the diagnosis of TOF with severe valvar and infundibular stenosis. There was an 11 mm subaortic non-restrictive VSD with equalization of right ventricular and left ventricular peak systolic pressures. There were multiple small aorto-pulmonary collaterals arising from descending thoracic aorta.

Informed consent was obtained from the parents before attempting total percutaneous correction, given

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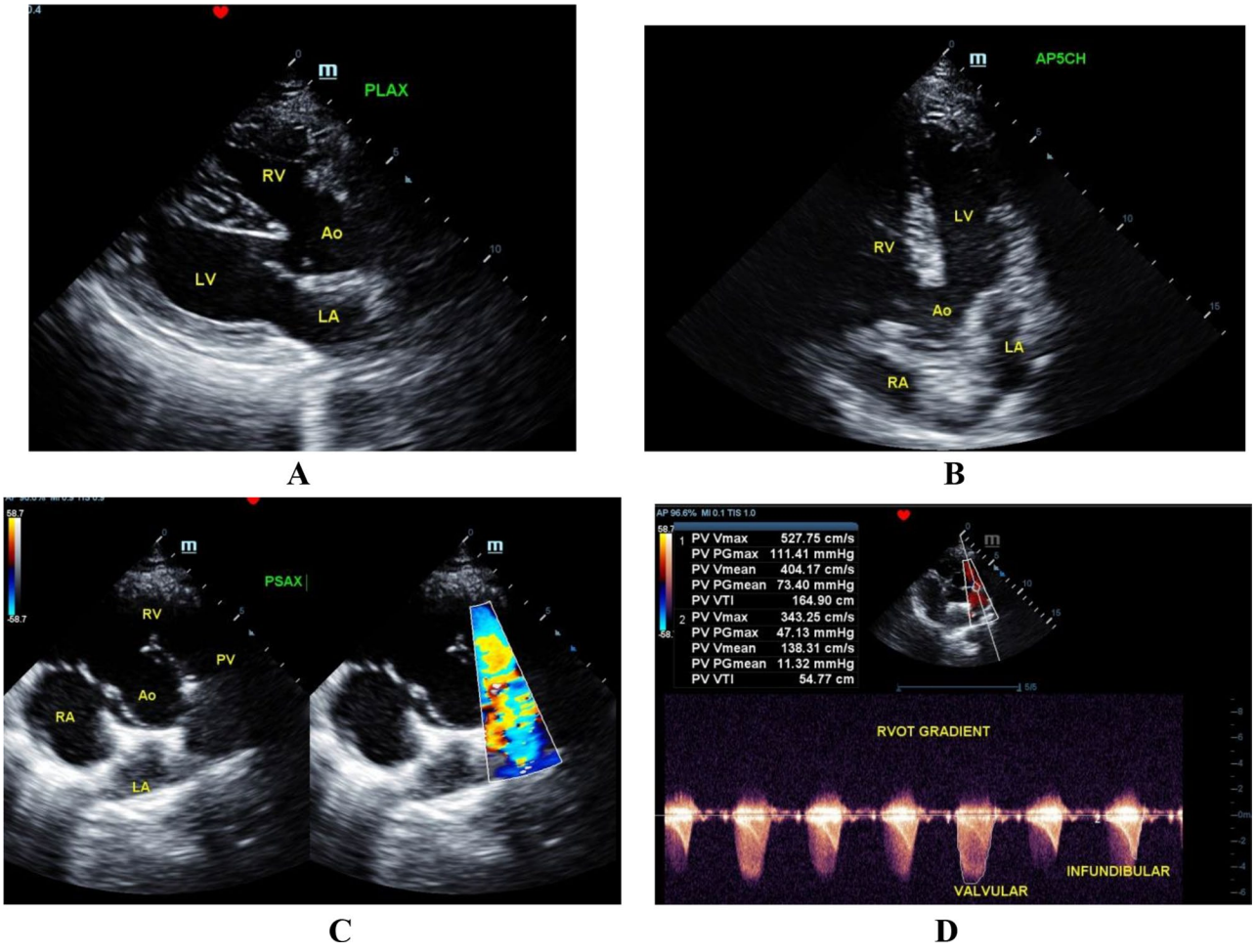


Fig. 1 Echocardiographic diagnosis of TOF. **A**) Parasternal long axis view showed VSD with aortic overriding, **B**) Apical 5 chamber view also showed the same features, **C**) Parasternal short axis view showed RVOT stenosis, **D**) Continuous wave Doppler recording of the RVOT gradient both at valvular and infundibular level

the parents and the patient were not willing to open heart surgery. Permission from the institutional review board was taken for this procedure.

In the 1st stage, we decided to address the RVOT obstruction. BPV was done to treat valvular pulmonary stenosis with the double-balloon technique and multiple dilatations until the waist of both the balloons disappeared at low pressures with two non-compliant Atlas Gold PTA dilatation catheters of 16×40 mm (Becton, Dickinson, and Company, NJ, USA) (Fig. 2A).

The peak instantaneous pressure gradient reduced from 90 mmHg to 50 mmHg. So, percutaneous closure of the conal arteries (in our case there were 2 conal branches, 1st we planned alcohol ablation of a larger branch) was planned. A simultaneous RV and right coronary artery angiogram clearly delineated two conal arteries that were supplying the large infundibular muscle. A temporary pacemaker implantation was done for back up support during the procedure and 48 h post-procedure for any brady-arrhythmic event. A 6 F JR3.5 was used to engage the right coronary artery. A 0.014" Floppy wire was placed distally in the larger conal artery. The branch was about 2.75 mm in diameter by quantitative coronary analysis (Fig. 2B). However, we did not have an over-the-wire balloon of the appropriate size. So, we used an alternative technique. A Finecross microcatheter (Terumo Medical Corporation, Japan) was advanced over the floppy wire. A 2.75×12 mm non-compliant balloon (Accuforce, Terumo Medical Corporation, Japan) was placed proximal to the tip of the microcatheter and was inflated at 12 atm pr. Injection of 1 ml contrast through the microcatheter confirmed the target area of ablation (infundibular muscle) and excluded any spillage of contrast to the main RCA (Fig. 2C). After confirming the assured target, 2 ml of absolute alcohol was injected through the microcatheter. the balloon was kept inflated for 1 minute to allow alcohol to reach the microcirculation. The patient complained of mild chest pain. Everything was removed after confirming the complete occlusion of the branch. The course of the smaller

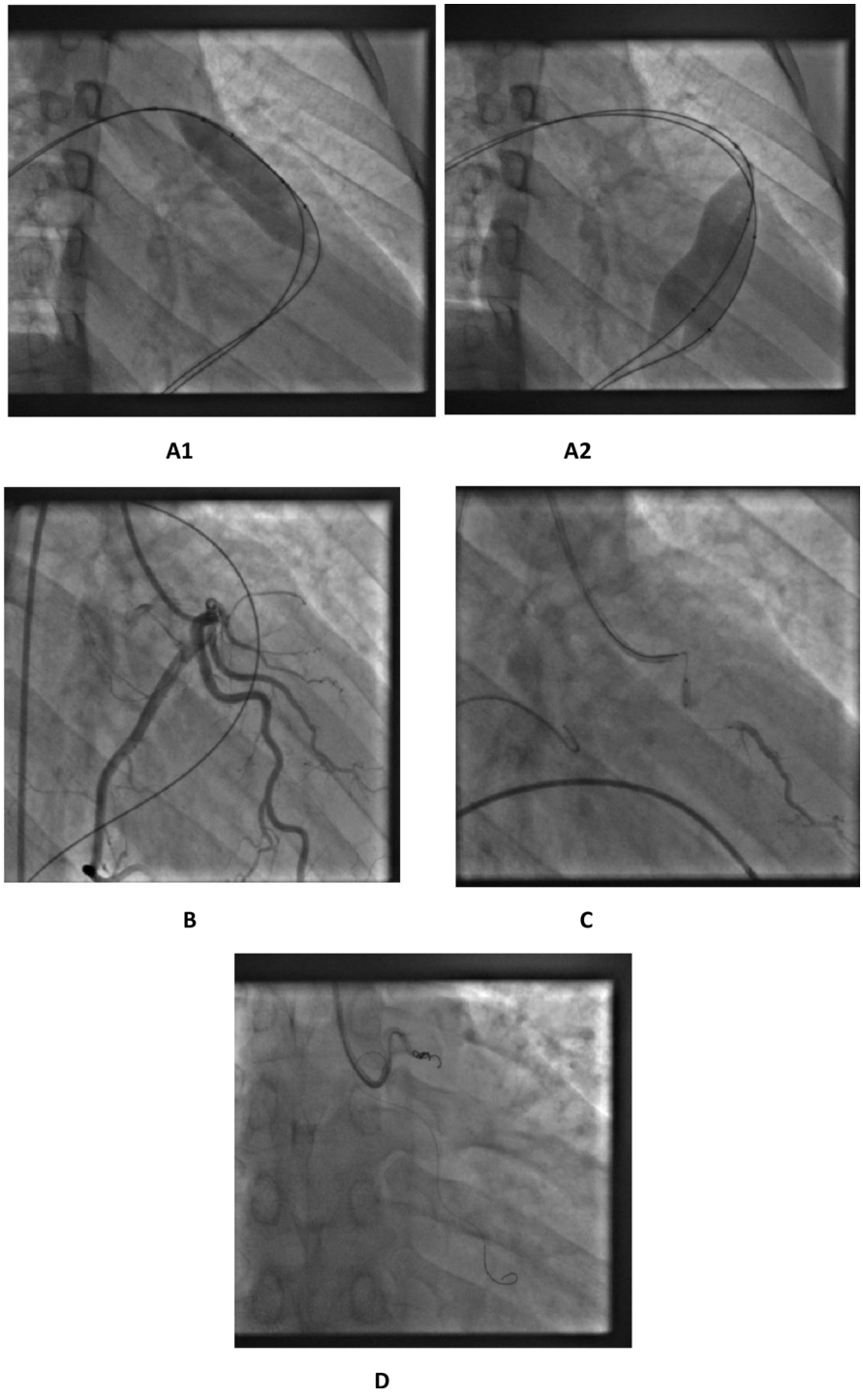


Fig. 2 Relief of RVOT obstruction. **A**) Fluoroscopic RAO view shows double balloon dilatation of RVOT (A1, Valvular; A2, Infundibular), **B**) Right coronary angiogram in RAO view showing two conal branches (**C**) Alcohol ablation of the large conal branch, **D**) Coil closure of the smaller conal branch

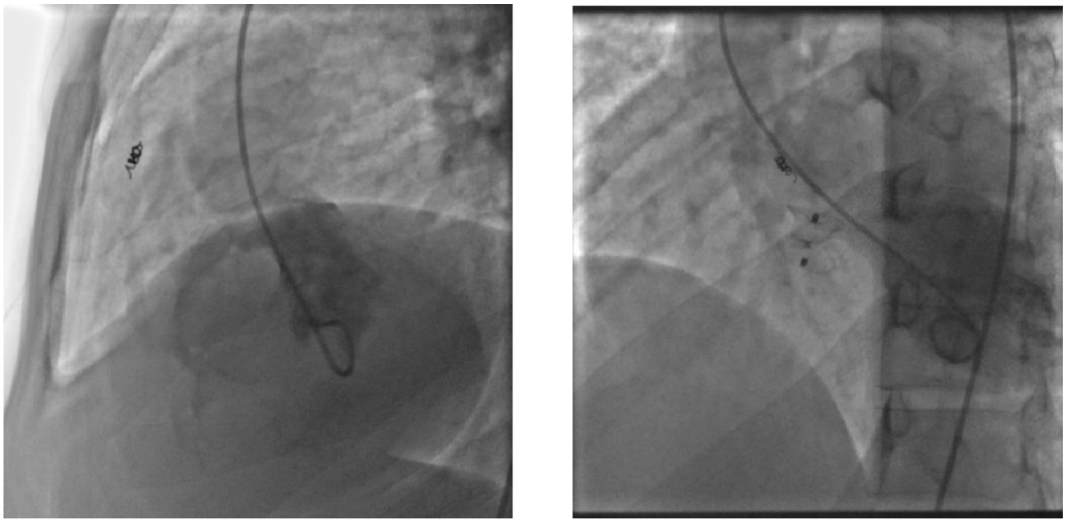
conal branch was unfavorable for the use of alcohol. So, we performed coil closure of that branch with two 0.018" coils (Hilal embolization microcoil, Cook, USA) through a Progreat microcatheter (Terumo Medical Corporation, Japan) to close it (Fig. 2D). There was a significant reduction of RVOT instantaneous pressure gradient from 90 mmHg (pre-procedure) to 20 mmHg. The patient's arterial oxygen saturation improved from 83 to 98% on room air. The patient had an uneventful hospital stay (the temporary pacemaker was removed on the 2nd day) and was discharged on 3rd day.

At 3 months follow-up, the patient was well, and the routine echocardiography showed an RVOT gradient of 30 mmHg with VSD with left to right shunt with a peak systolic pressure gradient of 60 mmHg. In the 2nd stage of correction, an LV angiogram was performed first (Fig. 3A). Then the VSD was easily crossed retrogradely from the left ventricle and an arteriovenous connection was established. A 7 French delivery sheath was taken from the venous side across the defect to the ascending aorta. A MFO Konar device 14−12 (Lifetech, China) was positioned across the defect. After confirmation by left ventriculogram (Fig. 3B) and transthoracic echocardiography, the device was released, with minimal residual flow through the device. Antegrade approach was used aligning the LV side of the device properly below the aortic cusp (discussed later). Post-closure transthoracic echocardiography showed minimal flow through the device across VSD (Fig. 3C) with 30 mm of Hg gradient across RVOT with arterial oxygen saturation being 98% on room air. At discharge he was prescribed Aspirin 75 mg once daily and Propranolol 20 mg twice daily. He is doing well after 4 months post-procedure. Echocardiography showed device in situ (Fig. 4A) with minimal flow, without any aortic regurgitation. There was no pulmonic regurgitation (Fig. 4B), estimated systolic pressure gradient across the RVOT was 25 mmHg (Fig. 4C).

Discussion

Surgical correction of TOF (ventricular septal defect patch closure and relief of RVOT obstruction; additional transannular patch in too small pulmonary annulus) provides excellent results in the current era. The characteristic abnormality in TOF is superior and anterior displacement of the infundibular septum resulting in a narrow and muscle-bound stenotic infundibulum, malaligned VSD, and aortic overriding with or without valvular pulmonic stenosis. Transcatheter total correction of TOF should include relief of RVOT obstruction and closure of VSD.

Pulmonary valve dilatation with specially curved valvulotome was first described by Brock in 1948 [4]. Balloon dilatation and RVOT stenting although used for relieving RVOT obstruction for many years did not give satisfactory results [5, 6]. Another way of dealing RVOT obstruction is to reduce septal muscle by initiating and creating necrosis. Ramakrishnan S. et al. had done alcohol ablation of the conal artery to reduce the infundibular muscle mass (as in hypertrophic obstructive cardiomyopathy) to relieve RVOT obstruction in a patient with infundibular stenosis [7]. Alcohol or coil occlusion of conal branch causes necrosis of the infundibular muscle thereby decreases RVOT obstruction. Alcohol appears to penetrate more deeply into the septal region and induces a larger area of necrosis, whereas coil or plug embolization leads primarily to ischemia and only secondarily to necrosis [8]. Sideris EB et al. performed transcatheter total correction in a patient of TOF variant with dominant pulmonary valvular stenosis by BPV followed by double balloon-patch (not available presently) repair of VSD [3]. In our case, we used the double-balloon technique to treat the valvular component of the RVOT obstruction and combined alcohol ablation and coil closure of two conal arteries to treat the infundibular component of RVOT obstruction. VSD in our case was closed with an MFO device. The double balloon technique provides a venting area between the balloons during inflation and thus ensures that the pulmonary valve orifice is not completely occluded, as it is by a single balloon [9].



A **B**

Fig. 3 Closure of the VSD. (**A**) LV angiogram showing VSD with a left to right shunt, (**B**) LV angiogram showing MFO device in position

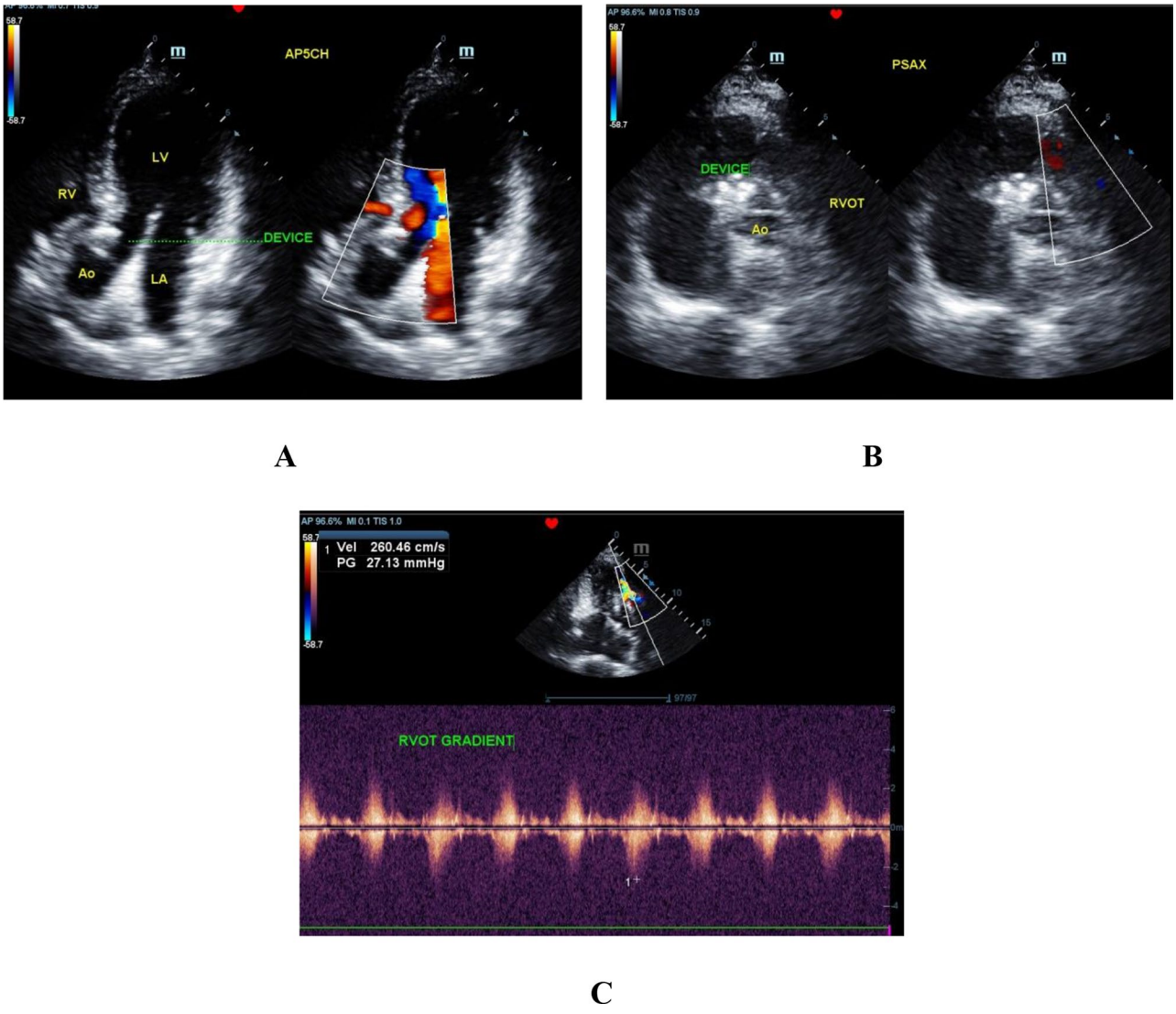


Fig. 4 Follow up echocardiographic findings. **A**) Apical 5 chamber view showed device in situ with minimal flow through the device without any aortic regurgitation; **B**) Parasternal short axis view showed no pulmonic regurgitation; **C**) Continuous wave Doppler recording of the RVOT

Non-compliant balloon maintains fixed diameter at high inflation pressure, offering more precise control over the balloon size. This is critically important to ensure accurate dilatation (uniform force distribution) of the stenotic pulmonary valve without overstretching the vessel causing injury [10]. With successful RVOT reconstruction small aorto-pulmonary collaterals regress slowly. However, larger (≥3 mm) collaterals require closure [11]. In our case the patient had multiple small collaterals which were not amenable for coil closure. With alcohol septal ablation in hypertrophic obstructive cardiomyopathy the LV outflow tract gradient response is triphasic, with immediate reduction, early reappearance, and by 3 months after procedure sustained fall [12, 13]. Expecting similar events in our patient we performed VSD device closure 3 months after the RVOT reconstruction. However, waiting 3 months carries a significant risk of increased pulmonary circulation and its consequences. So, this requires close monitoring.

MFO is a low profile double disc device with double sided screw [14]. LV side of the device has a disc with a highly conformable, flexible asymmetric lobe (similar to Amplatzer Duct Occluder 1) wider towards the disc. RV side of the device is a simple disc. We intended to position the device in a way that this lobe just hugs the ventricular side of the aortic cusp. As it is a very low profile device it should not cause significant aortic cup deformation (Fig. 5). So, we used antegrade approach in this patient.

To conclude, transcatheter total correction of TOF is feasible and effective in a selected group of patients who are surgically turned down. Whether the same technique

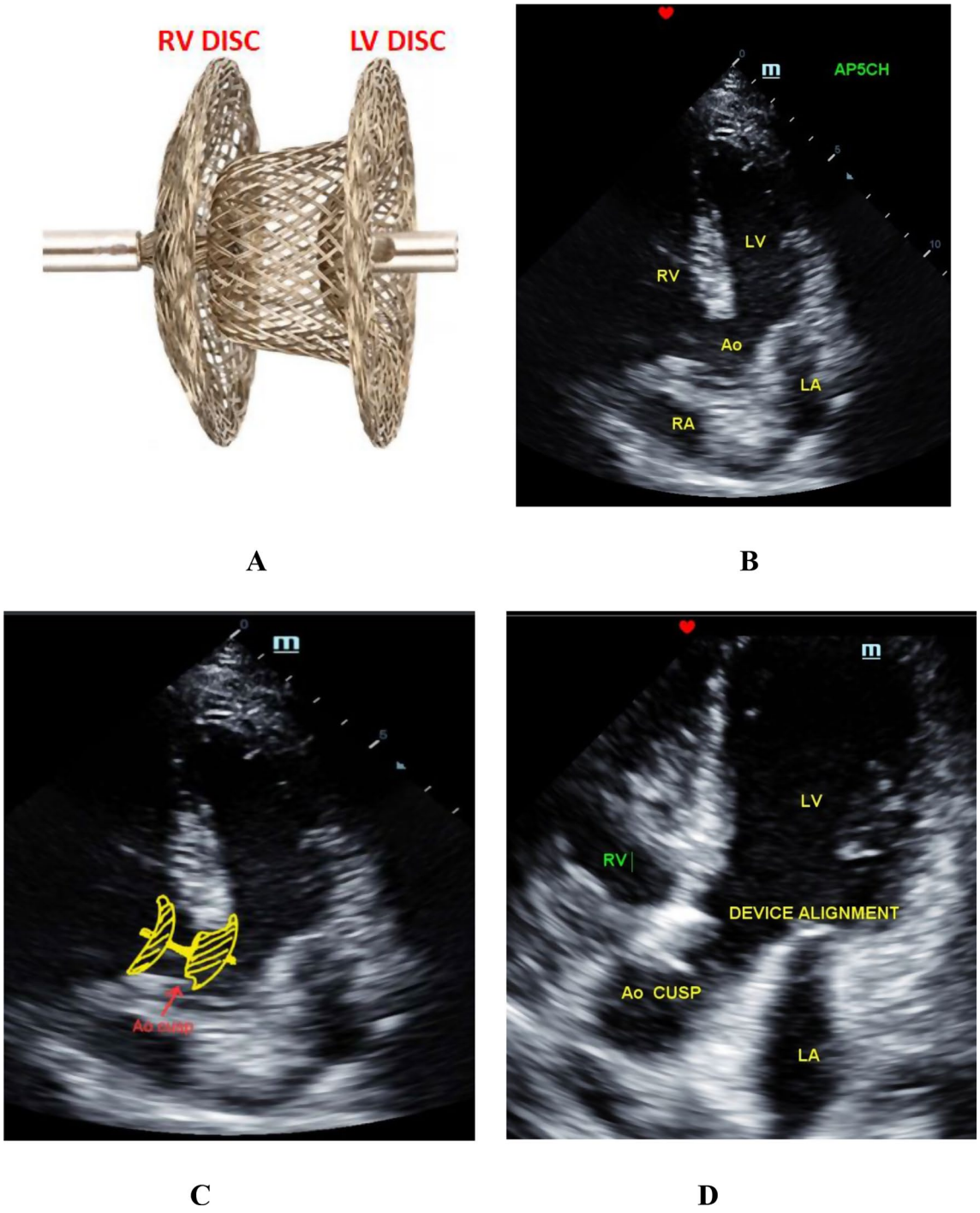


Fig. 5 MFO device alignment at the VSD. **A**) MFO device, **B**) Apical 5 chamber view showing the location of the VSD, **C**) Schematic diagram showing possible device alignment at the VSD, **D**) Device in situ

applies to the pediatric population is not clear to us. Moreover, the long-term risks of conal occlusion and future arrhythmia are unknown.

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s13019-024-03172-1.

Supplementary Material 1

Supplementary Material 2

Supplementary Material 3

Supplementary Material 4

Supplementary Material 5

Supplementary Material 6

Supplementary Material 7

Supplementary Material 8

Supplementary Material 9

Supplementary Material 10

Supplementary Material 11

Supplementary Material 12

Supplementary Material 13

Supplementary Material 14

Author contributions
A.D and G.L wrote the main manuscript D.K and L.M prepared Fig. 1 N.N.M and S.T prepared Fig. 3 All authors reviewed the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Disclosure
We have nothing to disclose.

Competing interests
The authors declare no competing interests.

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