Indian Journal of Thoracic and Cardiovascular Surgery (November–December 2023) 39(6):622–625
https://doi.org/10.1007/s12055-023-01564-x

CASE REPORT

Trans-aortic DORV repair

Amitabh Satsangi[1] · Hari Prasad[1] · Sheikh Mohd Murtaza[1] · V. Devagourou[1]

Received: 24 March 2023 / Revised: 23 June 2023 / Accepted: 26 June 2023 / Published online: 1 August 2023

Abstract
Double-outlet right ventricle (DORV) is a group of complex ventriculoarterial connections. In the literature, there are reports of trans-aortic correction of tetralogy of Fallot, as well as ventricular septal defects and complex DORV without right ventricular outflow tract obstruction. However, a pure trans-aortic approach for DORV ventricular septal defect (VSD) right ventricular outflow tract obstruction repair is not reported in the literature. We present a case of pure trans-aortic repair of DORV VSD pulmonary stenosis (PS) in an adult. A 20-year-old male with a known case of DORV, VSD, and PS presented with cyanosis. A pure trans-aortic repair of DORV was done. Complete trans-aortic DORV VSD PS repair is yet to be reported in the literature. The trans-aortic approach avoids a right atriotomy, right ventriculotomy, and injury to coronary arteries in cases of complicated tetralogy of Fallot and avoids injury to the tricuspid valve. This approach can be used in selected patients for intracardiac repair. The advantages are excellent visualization of the defects and avoidance of injury to the aortic cusps and bundle of His. In addition, placement of the VSD patch on the left ventricular outflow side may prevent residual shunts after repair.

Keywords Trans-aortic · DORV repair

Introduction

Double-outlet right ventricle (DORV) is a group of complex ventriculoarterial connections [1]. In most cases, the repair is performed via right atriotomy or a ventriculotomy [2]. In the literature, there are reports of trans-aortic correction of tetralogy of Fallot, complex DORV without right ventricular outflow tract obstruction (RVOTO) as well as trans-aortic closure of ventricular septal defects (VSD) [3, 4]. However, a pure trans-aortic approach for DORV VSD pulmonary stenosis (PS) repair is not reported in the literature. We present a case of pure trans-aortic repair of DORV VSD PS in an adult.

Case Report

A 20-year-old male with 1.52 m^2 body surface area (BSA) and known case of DORV, VSD, and PS presented with cyanosis. Echocardiography showed DORV with unrestricted VSD showing right to left shunting with occasional respiratory variation (bidirectional shunt), valvular and infundibular PS (gradient −90/44 mmHg), confluent pulmonary arteries with main pulmonary artery being 2.1 cm, pulmonary annulus size being 16.5 mm with Z score of −1.81, and McGoon ratio of 1.8. The right atrium and right ventricle were enlarged with normal biventricular function. Cardiac catheterization revealed a single large sub-aortic (SA)-VSD and normal ventricular function with a normal coronary pattern (Fig. 1). Severe valvular and sub-valvular PS was seen with confluent pulmonary arteries with normal sizes and a large significant aortopulmonary collateral (APC) requiring preoperative intervention was noted (Fig. 1A, B).

Post-coil embolization of the APC, the patient was electively taken up for intracardiac repair for DORV VSD and severe PS. Intraoperative trans-esophageal echocardiography showed a dense mass in the non-coronary sinus of the aorta, suggestive of a fresh thrombus. The etiology of the fresh thrombus was unknown and prior coil embolization was thought to be the incriminating factor. The trans-aortic approach was considered to address the thrombus in the non-coronary sinus. A transverse aortotomy was done, and a fresh thrombus was removed from the non-coronary cusp. The aortic leaflets were retracted and the sub-aortic VSD was visualized (Fig. 2A) (Video). Through the VSD, pulmonary valvotomy with sub-infundibular resection was done and RVOTO was relieved (Fig. 3). As per the Rowlatt

✉ Amitabh Satsangi
amoeba418@gmail.com

[1] Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi 10029, India

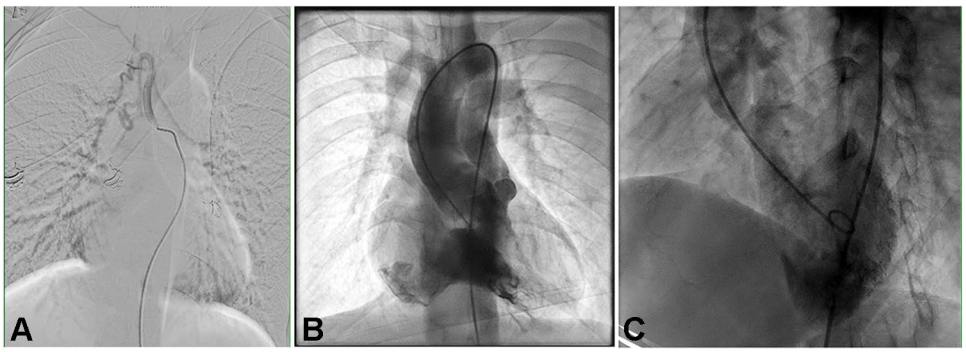

Fig. 1 Conventional angiogram. **A** APC coiling. **B** Preoperative double-outlet right ventricle. **C** Postoperative DORV repair with LV to aorta routing

chart [4], weight appropriate seventeen size Hegar dilators were passed serially across the right ventricular outflow tract (RVOT) which denoted an optimal relief of RVOTO. Trans-aortic VSD was visualized and the margins were identified. Adequate-size dacron patch was shaped and VSD repair was done with continuous 4′0 polypropylene suture. A saline test for the aortic valve showed no aortic valve incompetence. The aorta was closed in double layer (Fig. 2B). The patient was subsequently weaned off bypass with dobutamine 5 mic/kg/h. On intraoperative post-intra-cardiac repair trans-esophageal echocardiography (TEE), there was no residual VSD, minimal RVOT gradient of 30mmHg, and no features of left ventricular outflow tract obstruction. Routine closure was done. The postoperative course was uneventful. On 1-year follow-up, the patient is asymptomatic with New York Heart Association (NYHA) class 1. Follow-up echocardiography showed RVOT gradient of 20 mmHg with no residual VSD and good biventricular function. After 6 months of intracardiac repair, the patient had one episode of hemoptysis for which the patient underwent additional coiling of another APC successfully and a cardiac angiographic run revealed no residual VSD and RVOTO (Fig. 1C).

Discussion

DORV is an anomaly that is characterized by the great arteries arising from the right ventricle and associated with the VSD. In DORV and PS, due to restrictive pulmonary flow and a right to left shunt leading to an increased blood flow in the ascending aorta, the aorta is large, thus making the trans-aortic approach feasible.

A pure trans-aortic approach for total correction in tetralogy of Fallot has been reported earlier in which two cases were operated successfully [2]. Recently, trans-aortic approach has been used successfully in complex DORV with sub-aortic stenosis without any RVOTO [4]. Complete trans-aortic DORV VSD PS repair is yet to be reported in the literature. The trans-aortic approach avoids a right ventriculotomy and injury to coronary arteries in cases of complicated tetralogy of Fallot and avoids injury to the tricuspid valve. In DORV, since the VSD has a high location, trans-aortic approach provides a good view of all the margins of the VSD, thus providing excellent exposure [5, 6].

Trans-aortic approach in DORV VSD PS is recommended in doubly committed VSD, sub-aortic VSD, and VSD with sub-aortic stenosis, associated aortic regurgitation amenable to aortic valve repair or replacement, anomalous coronary artery anatomy, which contraindicates the conventional right atriotomy, and right ventriculotomy approach. This approach is preferred in patients with predominantly sub-valvular stenosis with mild to moderate valvular stenosis.

Contraindications for pure trans-aortic approach are sub-pulmonic and non-committed VSD, associated tricuspid valvular lesions, presence of a large atrial septal defect, and cases which require pulmonary valve reconstruction.

The intervening structure of the tricuspid valve and the high sub-aortic location of the VSD make the approach

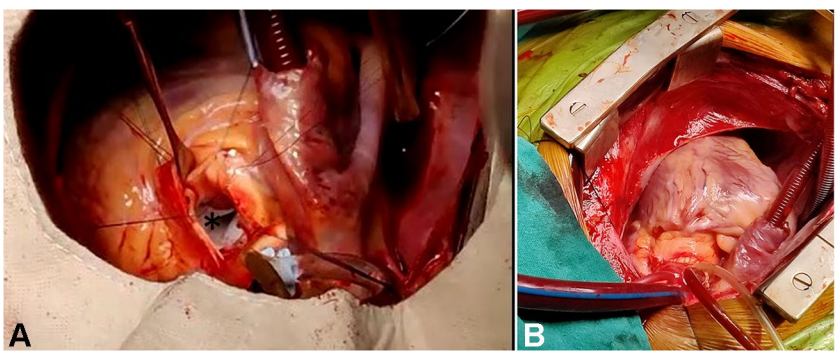

Fig. 2 Surgical view after opening the aorta. **A** VSD (asterisk) view through the trans-aortic approach. The aortic valve presented an overriding > 50%. The trans-aortic approach offered an excellent operative field to close the VSD. VSD, ventricular septal defect. **B** Trans-aortic post-DORV repair

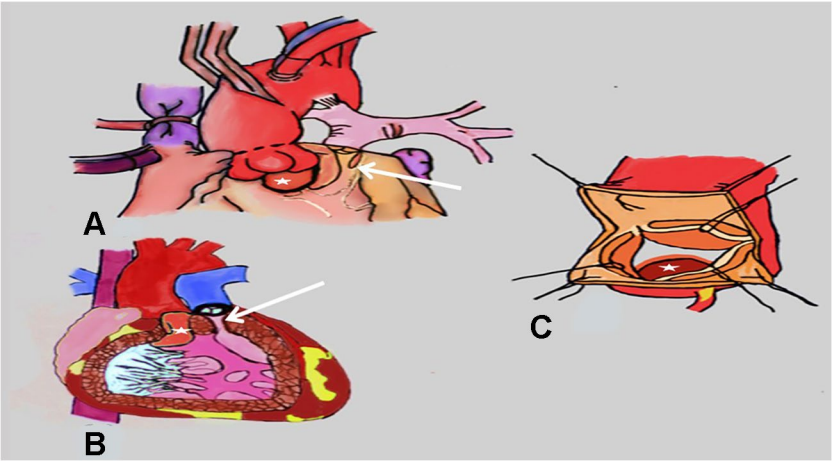

Fig. 3 Schematic representation. **A** Sub-aortic VSD (star) with right ventricular outflow tract obstruction—valvular and infundibular (arrow). **B** Trans-right ventricle view of sub-aortic VSD (star) and right ventricular outflow tract obstruction (arrow). **C** Trans-aortic view of VSD (star)

through the right atrium unsatisfactory. The approach through the aortic valve provides good exposure of the VSD. For many intra-cardiac surgical repairs, trans-aortic approach is well recognized.

Through the right atriotomy approach, sometimes it is difficult to size and shape the VSD patch. Under these circumstances, the patch tends to be bigger because a non-stenotic systemic route reconstruction is essential. When approaching through the aorta, the left ventricular outflow tract, including the VSD, is well visualized, and there is excellent access to a sub-aortic VSD. The infundibular obstruction is immediately downstream of the large VSD which is easily visualized and adequate resection can be done. Gentle traction on the aortic wall of the infundibular chamber brings the pulmonary valve annulus into view. Due to an exceptional visualization of the VSD, there is no fear of injury to the aortic cusps or the bundle of His [2]. In addition, arrhythmias that may occur after a trans-ventricular repair are entirely avoided.

The conduction bundle is located in the posterior and inferior rim of the VSD. No conduction abnormality was noted in the above patient. Careful tailoring of the patch to avoid constriction of the VSD, which was the outlet of the left ventricle, prevented obstruction of the left ventricle outflow. The tunnel made through aortotomy is narrower than those made through a right ventriculotomy and reconstruction of the RVOT without extracardiac conduits. The trans-aortic approach also provides the opportunity for assessment of the left ventricular outflow tract under direct vision, thus decreasing the chances of left ventricular outflow tract obstruction during VSD closure (Fig. 2).

The advantages of the trans-aortic approach in systemic intraventricular reconstruction lead to improved geometric and hemodynamic results in patients with a small ventricular cavity. The disadvantages of a trans-aortic approach include prolonged aortic cross-clamp time and potential traction or trauma of the aortic valve. In this patient above described, the aortic root and VSD were large. There was no fear of injury to the aortic cusps or the bundle of His because visualization of VSD was exceptional.

Conclusion

This approach has its unique benefits in well-selected cases for intra-cardiac repair for DORV. The advantages are excellent visualization of the defects and avoidance of injury to the aortic cusps and bundle of His. In addition, placement of the VSD patch on the left ventricular outflow side prevents left ventricle outflow tract obstruction.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s12055-023-01564-x.

Funding None.

Declarations

Ethical approval Not applicable.

Informed consent Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Human rights Not applicable.

Conflict of interest No conflict of interests.

References

1. Oladunjoye O, Piekarski B, Baird C, Banka P, Marx G, Del Nido PJ, et al. Repair of double outlet right ventricle: midterm outcomes. J Thorac Cardiovasc Surg. 2020;159:254–64.
2. Kumar AS, Talwar S, Velayoudam D. Transaortic correction of tetralogy of Fallot and similar defects. J Thorac Cardiovasc Surg. 2006;131:1185–1186.e1.
3. Harlan BJ, Cooley DA. Transaortic repair of double-outlet right ventricle with situs versus, 1-loop, 1-malposition (I, L, L), subaortic ventricular septal defect, and associated anomalies. J Thorac Cardiovasc Surg. 1976;72:547–52.
4. Rowlatt UF, Rimoldi HJA, Lev M. The quantitative anatomy of the normal child's heart. Pediatr Clin North Am. 1963;10:499–588.
5. Fernandez-Doblas J, Pamies-Catalan A, Dolader P, Ferrer Q, Abella RF. Usefulness of transaortic approach for a complex double-outlet right ventricle. Indian J Thorac Cardiovasc Surg. 2022;38:84–6.
6. Sakamoto K, Charpentier A, Popescu S, De Geeter B, Eisenmann B. Transaortic approach in double-outlet right ventricle with subaortic ventricular septal defect. Ann Thorac Surg. 1997;64,856–8.