

Left Sided Surgical Approach in Dextrocardia DCRV

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Abstract: *Introduction:* Double Chamber Right Ventricle (DCRV) and dextrocardia are two congenital cardiac conditions with rare incidence. A combination of both these condition is extremely rare with only one reported case to the best of our knowledge. *Case:* 8 year old patient diagnosed with DCRV dextrocardia successfully underwent intra cardiac repair at our institute. *Background:* We present a case report of this case, giving a description of its anatomy and our surgical approach to it. We give an account of our unconventional left sided approach to it. *Conclusion:* Left sided approach in dextrocardia is an easier approach and gives a better visualization of VSD in DCRV.

1. Introduction

Dextrocardia is a rare cardiac malposition, often associated with multiple cardiac anomalies. Surgery in dextrocardia patients is rarely encountered by low volume centres and poses a significant challenge for the surgical team. It requires a prospective strategy and reorientation for cannulation, approach to intra-cardiac lesion and associated anatomical variations. DCRV in a setting of dextrocardia is extremely rare with limited surgical understanding. We present the anatomical description and our surgical approach in a paediatric case of DCRV with subaortic ventricular septal defect (VSD) and subaortic spur with dextrocardia situs inversus.

2. Case Summary

8 year old acyanotic boy hailing from rural India, presented with effort intolerance and repeated lower respiratory tract infections since 3 months of age. Heart rate was 90 per minute, blood pressure 110/70 mm Hg with a saturation of 97%. Pan systolic murmur in right parasternal region radiating all over the right chest wall was heard. Chest X-ray showed dextrocardia and situs inversus. Two-dimensional echocardiography confirmed dextrocardia with situs inversus. DCRV with hypertrophied muscle bundle in the right ventricular outflow region with peak gradient of 69mm Hg was diagnosed. A 2X1cm non-restrictive sub-aortic VSD with left to right shunt and a sub aortic spur 6 mm in size were also identified. Cardiac catheterization confirmed the diagnosis.

Surgery was performed on conventional cardiopulmonary bypass with aorto-bicaval cannulation, moderate hypothermia and delnido cardioplegia with right handed surgeon operating from the left side and pump placed behind him (Figure 1A, 1B). The patient was placed on the left edge of the operating table. Standard midline sternotomy, pump tubing fixation, pursestring and cannulation were performed from the right side for its convenience as per protocol surgical steps.

Pericardiectomy was performed near the midline leaving a smaller patch on the morphological right atrial (RA) side for a better exposure. Heart was occupying right hemithorax

with apex directed towards right. Morphological RA with appendage was on the left towards midline and received both caevae. The morphological left atrium and appendage were posteriorly; receiving all the four pulmonary veins. Right Ventricle (RV) was the anteriorly placed ventricle giving rise to normally branching pulmonary artery. RV showed dimpling below outflow tract suggesting DCRV. Aorta was to the left and anterior to pulmonary artery, left sided arch with normal branching. Large branches of Right coronary artery were seen over right ventricle including prominent conal branches over right ventricular outflow tract (RVOT). RA was opened parallel to atrio-ventricular groove by an incision placed to the left of RA appendage. RA stay sutures were placed near the caudally directed tricuspid valve annulus (Figure 2A) and 2x1 cm sub-aortic VSD could be visualized through it. Hypertrophied RVOT band divided the ventricle to proximal and distal chambers of DCRV which was excised from an incision in RVOT. Hegar which was sized adequately for the surface area could be passed from the right ventricle, across the excised band into RVOT and pulmonary artery comfortably. (Figure) Pulmonary Valve and pulmonary arteries were normal. Sub-aortic spur was visualized below the aortic leaflets and excised circumferentially. VSD closure was done through the RA using a SFD patch in interrupted horizontal pledget reinforced mattress manner starting from the tricuspid annulus, proceeding towards the aortic rim and continuing to the postero-inferior margin. Conduction tissue was assumed to be occupying the infero-posterior margin of VSD and caution was taken to stay away from it. The cardiac chambers were closed and the patient was weaned off cardio pulmonary bypass (CPB) uneventfully. Decannulation was performed from the right side. Post operatively patient was in normal sinus rhythm, RVOT gradient of 25mm Hg, no aortic regurgitation, no shunt across VSD and discharged uneventfully.

3. Discussion

DCRV is a rare congenital heart defect in which anomalous hypertrophied muscle bands running from ventricular septum inferior to the insertion of the septal leaflet of the tricuspid valve to the anterior wall of the RV dividing RV into a proximal high pressure sinus portion and a distal low pressure infundibular chamber. The bands may be due to localised growth of the trabeculated myocardium early in

development or arrested incorporation of the primitive bulbus cordis into the RV body.¹

DCRV is a rare congenital anomaly constituting 1-2.6% of congenital heart disease.² Situs inversus accompanied with dextrocardia is also a rare occurring in 0.01% of live births. DCRV with dextrocardia situs inversus has been reported only once.³ DCRV may have coexisting VSD (80-90%), subaortic stenosis, pulmonary valve stenosis, double-outlet right ventricle, Tetralogy of Fallot, or anomalous pulmonary venous drainage, among others. Thus DCRV dextrocardia situs inversus subaortic spur contributing to left ventricular outflow tract obstruction becomes extremely rare.

Diagnosis of the condition is an indication for corrective intervention. Surgical resection of the bundle, as well as repair of other anomalies is usually indicated as soon as the diagnosis is made.⁴

We considered performing the steps till arresting the heart on conventional aorto-bicaval CPB to be performed from the routine right side for its convenience as protocol for all members of the surgical team including perfusionist and scrub nurse. The tubings were placed as routine. Left sided surgical approach was considered appropriate for intracardiac repair of VSD as the intracardiac anatomy and conduction bundle was assumed to be mirror image.

Orientating the surgical team with this plan was an important contributor to the smooth performance of surgery.

Open heart surgery in dextrocardia is technically demanding and the surgical team is required to make certain adjustments to their surgical techniques. Only anecdotal reports of surgical techniques in dextrocardia are available. In cases of dextrocardia, the cannulation for CPB is difficult because the venae cavae and RA are situated more posteriorly than normal.⁶ This has been made relatively easy by operating from the left side of the patient as was done in our case. The left side approach also gives a good visualization of VSD.

In conclusion, cardiac surgery in a patient with dextrocardia requires a good surgical planning. The pathology and

anatomy has to be defined clearly and operative problems need to be anticipated.

4. Acknowledgments

None

5. Classification

CHD, Dextrocardia; CHD, DCRV VSD; Subaortic spur

6. Funding Issues/ Conflict Of Interest/ Competing Interest

None

7. Prior Related Publication/Presentation

None

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