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Title of the article: Bi-ventricular repair of Double Outlet Left ventricle - Experience and Review of Literature

Running title: Double outlet left ventricle - Case series

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Abstract:

Background:

Double-outlet left ventricle (DOLV) is a rare congenital cardiac anomaly. The aorta and the main pulmonary arterial trunk arises predominantly from the left ventricle and is associated with a malaligned ventricular septal defect(VSD), various degrees of hypoplasia of the right ventricle, and presence or absence of pulmonary stenosis. Bi-ventricular repair is the preferred treatment option whenever possible. Multiple techniques for bi-ventricular repair have been described. The best option for DOLV correction is by translocating the pulmonary root from the left ventricle(LV) to the right ventricle(RV).[1] In this series, we report four patients who underwent biventricular repair of DOLV with excellent outcomes.

Methods:

This is a retrospective study of four patients who underwent surgical correction of DOLV between January 2014 and December 2018. We collected all patient details from the institute patient record system. Echocardiographic data were obtained from the records. Intraoperative charts were reviewed for further information on the surgical procedure and cardiopulmonary bypass. Postoperative data included survival, functional status, and followup echocardiography.

Results:

Of the four children, three underwent pulmonary root translocation, and one child underwent a Reparation al etage Ventriculaire(REV) procedure. There was no mortality in our series, and all children are in a stable clinical condition in the recent follow-up, and no re-operations or interventions were required following primary surgical correction.

Conclusion:

DOLV is anatomically and surgically a challenging subset. Pulmonary root

translocation in this anatomy is technically challenging but safe and superior option when compared to other alternative surgical procedures. Pulmonary root translocation can be performed with excellent results, even in infants.

Keywords:
Congenital heart disease, Double outlet left ventricle, Pulmonary root translocation

Introduction:

Double-outlet left ventricle(DOLV) is a rare anomalous ventriculoarterial connection with both aorta and the pulmonary artery arising entirely or predominantly from the morphologic left ventricle.[1] The relationship of the great arteries, location of the VSD, and the presence of tricuspid valve abnormalities are important morphological factors to be considered before surgical repair of DOLV. There are various surgical options for biventricular repair for DOLV, which include Arterial Switch Operation(ASO), Rastelli repair, and Pulmonary root translocation.[6] In the presence of tricuspid valve abnormalities and associated hypoplasia of the right ventricle, the univentricular repair is the other alternate option.

Materials and Methods:

Four patients who underwent surgical repair of DOLV between January 2014 to December 2018 were included for the retrospective analysis. We collected the pre-operative, intra-operative, and postoperative data from the patient records. All patients were followed up with recent follow-up echocardiography. The following morphological, anatomical details of all four patients were noted, namely, the relationship of the great arteries, the position of the ventricular septal defect (VSD), the coronary anatomy, the morphology and size of the tricuspid valve, and the right ventricular size(Table 1). The intraoperative and postop-

erative details were recorded. The condition is rare. Hence only a descriptive analysis has been done in this study along with a review of the literature.

All patients were approached via standard median sternotomy. Aorta and the pulmonary artery was mobilized up to the hilum. Cardiopulmonary bypass(CPB) was initiated after high ascending aortic and bicaval cannulation. PDA was doubly ligated and divided. LA vent was inserted. We used antegrade cardioplegic arrest with Del Nido cardioplegia in all the patients. Right atriotomy made, and intracardiac morphology assessed. After confirming it to be DOLV, the pulmonary root was harvested, taking care not to injure the coronaries, and the defect in the LV was closed with a bovine pericardial patch. Through the trans right atrial approach, intracardiac tunneling of LV to aorta was done using the Goretex patch. RVOT incision made, and the pulmonary root was anastomosed to the RVOT, anteriorly augmented with a bovine pericardial patch. Patient 3 underwent REV procedure as the pulmonary valve was not suitable for root translocation. Figure 1 shows the schematic representation of pulmonary root translocation.

Results:

The median age at presentation was 19 months (1 month to 12 years), with males and females equally distributed in our series. The median weight of the children at presentation was 6.75 kg ranging from 2.6 kg to 26 kg. All four patients were cyanotic at presentation. Two patients were diagnosed as Dextro Transposition of great arteries with VSD, and the other two patients were diagnosed as Double outlet right ventricle with VSD and severe pulmonary stenosis by pre-operative echocardiography. Intra-operative assessment of the anatomy confirmed the diagnosis of DOLV in all four patients.

Morphologically, the atria and the viscera were in situs solitus in all four patients. All patients had a side by side great arterial relationship. The aorta was to the right of the pul-

monary artery or slightly posterior. The VSD was large and subaortic, and the tricuspid valve was normal with a normal 'Z" score. The Right ventricle in all four patients was adequate in size, thus committing the patients for a bi-ventricular repair. Three patients had abnormal coronary anatomy. Table 1 shows the morphology and coronary anatomy of individual patients.

All the four children underwent bi-ventricular repair that includes pulmonary root translocation in three patients and REV procedure in one. There was no mortality in our series. The cross-clamp time and cardiopulmonary bypass time of all the four patients are shown in table 1. In patient 1 and patient 4 with a preoperative diagnosis of dTGA, an arterial switch was not done due to the presence of moderate pulmonary incompetence and dilated pulmonary annulus, while patient 2 had significant infundibular stenosis. All three patients underwent Pulmonary root translocation with VSD closure. Patient 4 had significant annular stenosis with a dysplastic pulmonary valve and hence underwent REV procedure with a monocusp reconstruction of RVOT due to the unavailability of appropriately sized homograft.

All children were discharged in stable condition and are on regular followup. Ross modified score[21] was used to assess the functional status in the follow-up period, which was found to be less than two in all children, and the last follow-up echocardiography showed good LV function and no significant left ventricular or right ventricular outflow tract obstruction (Table 2).

Discussion:

DOLV has an incidence of less than 1 in 200,000 live births[2]. Both the great arterial trunks arise either entirely or partly from the morphological left ventricle. A variety of lesions are associated with DOLV, namely, pulmonic or subpulmonic stenosis (or both), tri-

cuspid value anomalies, and right ventricular hypoplasia.[3,4] There are various hypothesis that explains the embryogenesis of this rare condition. Paul et al. stated that anomalous differential growth of conal septum[5] beneath the semilunar valves, as the most contributing factor in the development of the relationship(both normal and abnormal) between the great arteries. Anderson et al.[6] proposed that malabsorption or malrotation of the sub arterial portion of the ventricular septum that separates the RV infundibulum from both great arteries leads to the development of DOLV.

Bharati et al.[7] have described and classified DOLV. The relationship between the great arterial trunks and the position of the VSD is of prime importance in understanding this complex anatomy. The great arterial relationship in DOLV is either side by side with aorta to the right or right and posterior to the Pulmonary trunk or with an anteroposterior relationship with aorta anterior to the pulmonary trunk or slightly to the right or left. All four patients in our series had side by side relationship of great arteries with the aorta to the right of the pulmonary trunk in all patients.

The VSD in this rare anatomy is also heterogeneous in its location. It is usually subaortic but can also be subpulmonic or large VSD overriding both great arterial trunks. Remote VSD has also been reported in this anatomy. All four patients in this series had a subaortic VSD. Due to the heterogeneous relationship of the great arterial trunk and the VSD, the diagnosis can often be confused with dTGA or DORV, VSD. In our series too, preoperatively, patients 1 and 4 were diagnosed as dTGA, VSD, and patients 2 and 4 were diagnosed as DORV, VSD with pulmonary stenosis.

Depending on the size of the right ventricle and tricuspid valve, along with the position of the VSD, DOLV is usually amenable to bi-ventricular repair.[7] Based on the hypoplasia of the right ventricle, DOLV is classified as type 1 and type 2. In type 1 DOLV, the

RV is normal and hence suitable for a bi-ventricular repair. Type 2 DOLV, the RV is hypoplastic and goes for a univentricular repair.[8] All four patients in this series were type 1 with good sized right ventricle.

The relative position of the VSD to the great arteries and the presence or absence of pulmonary stenosis is the prime determinant in deciding which type of surgical procedure for a bi-ventricular repair.[9] The classical surgical approach for bi-ventricular repair is the Rastelli procedure.[10,11] This procedure involves intracardiac baffling with right ventricle to pulmonary artery conduit. This technique may not be suitable when the VSD is restrictive or remote or when there is a prominent subaortic conus. The patients also will eventually outgrow the RV to Pulmonary artery conduit requiring replacement of the conduit.[11]

In the absence of pulmonary stenosis or when the stenosis is infundibular and resectable, intracardiac baffling of LV to pulmonary artery followed by Arterial switch is an acceptable procedure.[12] The drawback of this procedure is the need for coronary transfer, which is especially tricky when the great arterial relationship is side by side, and the abnormal coronary pattern is present. 3 of the 4 patients in our series did not have a normal coronary pattern (table1). The presence of a dilated pulmonary root and pulmonary incompetence is also a relative contraindication for this procedure. The other alternate options in the absence of pulmonary stenosis are intracardiac baffling when the VSD is subpulmonic and the use of a boomerang-shaped patch when the VSD is subaortic in location.[13]

VSD closure, along with pulmonary root translocation onto the right ventricle for DOLV, was proposed by Chiavarelli and associates in 1992.[14] There is only a limited literature review on pulmonary root translocation for DOLV, as it is a rare presentation (table 3). Three patients in this series underwent pulmonary root translocation. When the pulmonary artery lies much posteriorly relative to the aorta, the surgical options include ASO or

pulmonary root translocation, but the preferred procedure would be pulmonary root translocation[14,15] as performed in our centre. Pulmonary root translocation involves dissection and mobilization of the main pulmonary artery along with the pulmonary valve. Intraoperative assessment is essential, and if the subpulmonic conus is absent, the pulmonary root will be in continuity with the atrioventricular valve. Mobilization of the pulmonary root should be done very carefully without damaging the atrioventricular valve in this scenario. The harvesting of the pulmonary root is also technically challenging in the presence of double looping coronary anatomy. One patient in this series had double looping coronary anatomy. [16,17,18]

The primary advantage of pulmonary root translocation is that coronaries are not mobilized, which is beneficial, especially when the coronary pattern is abnormal. 3 out of the 4 patients in this series had an abnormal coronary pattern. The reconstruction of the right ventricular outflow in this procedure involves the use of native tissue along with the pulmonary valve. This preserves the growth potential of the reconstructed RVOT.[19] These factors lead to a decreased likelihood of re-operation. The more frequently reported late complications of the arterial switch, namely aortic insufficiency, pulmonary stenosis, and coronary lesion, are also avoided in this technique[16,17,20]. We believe that patients undergoing pulmonary root translocation for DOLV will have a better quality of life index when compared to other alternative procedures. Two of the three patients in our series were infants. All the patients at follow-up continued to remain in good clinical condition.

Conclusion:

DOLV is anatomically and surgically a challenging subset. Pulmonary root translocation in this anatomy is technically challenging but safe and superior option when compared

to other alternative surgical procedures. Pulmonary root translocation can be performed with excellent results, even in infants.

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