INTRODUCTION

Double-outlet left atrium (DOLA) is an extremely rare anomaly; a recent report found that only seven cases had been described so far.[1] To our knowledge, there have been only two published reports of repair of DOLA.[2,3] From the Indian subcontinent, only two such cases have been reported, neither of which was operated upon for different reasons.[1,4] We here describe the clinical detail, echocardiographic features, angiographic evaluation, and surgical repair of DOLA with intact ventricular septum and common atrioventricular (AV) valve in a 2-year-old boy.

CASE REPORT

The patient was a 2-year-old boy born out of nonconsanguineous marriage from Iraq. He had an unremarkable perinatal history and was breastfed till about 6 months of age which was associated with forehead sweating and suck-rest-suck cycle. Bluish discoloration of lips was noted by his parents at around 4 months of age and evaluation at a local facility suspected a congenital heart disease for which he was referred to our center. His physical examination revealed a heart rate of 75 beats/min, blood pressure of 108/66 mmHg, and respiratory rate of 34/min with oxygen saturation of 85%–87% in room air. An ejection systolic murmur grade 3/6 at the left upper sternal border and a normal split second heart sound were heard on auscultation. Electrocardiogram showed left atrial enlargement, with left-axis deviation, without ventricular hypertrophy. Chest radiograph showed cardiomegaly and increased pulmonary vascularity. A transthoracic echocardiography (ECHO) [Figure 1a, b] revealed situs solitus, levocardia, and d-loop ventricles with normal pulmonary and systemic venous drainage. There was right atrial outlet atresia, with the only outlet for the right atrium (RA) being a large secundum atrial septal defect (ASD). The LA emptied into the right and left ventricles through two components of common AV valve. There was mild AV regurgitation. The great vessels were normal in origin and position.

The disconcerting part was the right ventricular (RV) inflow which was estimated to have a Z-score of −2 [Figure 1a, b].
He was scheduled for a cardiac catheterization study primarily to measure the pulmonary artery pressures and pulmonary vascular resistance so as to assess suitability for repair. In addition, a visual impression of RV volume and size was also helpful, as on ECHO, the RV inflow and RV were small for weight. Cardiac catheterization revealed normal pulmonary artery pressures and adequate-sized RV.

After evaluation, a decision was taken to proceed for a possible intracardiac repair. One and a half ventricle repair was kept as the second option.

Inspection of the inside of the RA revealed normally located caval openings, a 20-mm secundum ASD, a large coronary sinus, and no AV connection. The atrial septum was excised completely and LA was exposed. The pulmonary venous openings and left appendage were positioned normally. There was a common AV valve with bridging leaflets attached to ventricular septal crest. There was no ventricular septal defect (VSD).

The right component of the valve was smaller and resembled a morphologic tricuspid valve with cords attached to the ventricular septum and papillary muscles in morphologic RV, and the left component had a cleft which was moderately regurgitant on saline testing and it resembled a morphologic mitral valve with its tensor apparatus supported by two papillary muscles only. There were no cords crossing the ventricular septum to suggest straddling.

The cleft was approximated with two interrupted polypropylene (Ethicon LLC, San Lorenzo, PR 00754, USA) mattress sutures. A redundant Dacron (Bard Shannon Limited, Humacao, Puerto Rico 00791) patch was used to separate the two AV orifices and to create a new atrial septum committing systemic veins to tricuspid and pulmonary venous return to mitral valve. To allow for an adequate pathway, the coronary sinus was cut back into the LA generously so as to deflect the patch into voluminous LA. Coronary sinus was eventually dedicated to RA. After septation, the constructed pathway accepted an Hegar dilator of 19, against an expected size of 20. The patient was weaned off cardiopulmonary bypass with the consideration of adding a cavopulmonary shunt depending on the hemodynamic and intraoperative echocardiographic assessment. The weaning was uneventful with central venous pressure in single digit, ECHO revealed excellent ventricular function with some flow acceleration in the tricuspid inflow (a mean gradient of 4 mmHg); there were two tiny jets of mitral regurgitation.

Postoperative course was uneventful. Predischarge ECHO [Figure 1c, d] revealed good flow across tricuspid component (mean pressure gradient of 3 mmHg, heart rate = 81/min), mild tricuspid regurgitation, mild mitral regurgitation, ASD patch in situ with no residual shunt, and normal ventricular function. In the follow-up, the child has performed well with good exercise tolerance.

**DISCUSSION**

Double-outlet atrium is a very rare cardiac anomaly in which one of the atriums connects to both ventricles. In DOLA, the LA drains into both ventricles and the right atrial outlet is just an ASD. The nomenclature and structure of the AV valve in this setting can be variable.

It was Van Mierop et al. who provided its initial description as a variant of the so-called “endocardial cushion defect” in the setting of common AV junction; in this setting, the extreme malalignment of the atrial septum relative to the muscular ventricular septum results in double outlet from either the LA or the RA. They assumed that in DOLA, the left AV valve is a common AV valve and the abnormality results from the extreme rightward malalignment of the atrial septum which fuses with the right margin of the AV canal, causing right atrial outlet atresia with LA draining into both ventricles.

Kirally et al. published their findings on the morphologic features of uniatrial but biventricular connection. They theorized that the anomaly with a striking similarity with DOLA was produced as a result of the atresia of the right AV valve and straddling of the left AV valve. However, all the specimens described in their report had VSD of varying size, allowing a straddling to happen. In addition, the anterosuperior leaflet was typically free floating and the principal line of coaptation of the bridging leaflets was always perpendicular to the
plane of the ventricular septum which did not permit the description of valve as either tricuspid or mitral. The leaflets were dysplastic with short cords and fused attachments of the crest and the valve per se had a propensity to failure toward regurgitation. There were sufficiently severe valvar and subvalvar anomalies not to allow a successful biventricular repair. They suggested that the best course possible for such patients was a single ventricle pathway.

More recently, Praagh[7] very convincingly put forth the concept that double-outlet atria were a rare form of ventriculo-atrial malalignment defects. Embryologically, during the 5th week of fetal life, the ventricular septum moves leftward beginning to become aligned with the atrial septum and the tricuspid valve now opens into the RV, thus the concordant alignment is RA to RV and LA to LV. DOLA represents a defect in ventriculo-atrial alignment, wherein the ventricles and ventricular septum have moved too far to the left relative to the atria and the atrial septum; consequently, the LA opens into both the LV and the RV and the RA opens into nothing because the RV lies to the left of the RA. As per this schema, the atria and the AV valves are essentially normally formed. The anatomic finding of the present case can be best explained by the Van Praagh model. In this setting, an excision of the malaligned atrial septum and reorientation of this with a patch placed to redirect the venous return to their appropriate ventricles are eminently possible, resulting in biventricular repair.

Given the wide spectrum of the anatomic subsets, all resulting in a uniatrial but biventricular arrangement, it is imperative that a systemic stepwise approach of segmental analysis of three cardiac segments and two junctions be undertaken to clarify the treatment. In a setting like this, if the two AV valves are phenotypically normal and straddling is absent, a biventricular approach should be attempted.

In our opinion, the atrial septal patch should be kept a little redundant to give volume to the reconstructed RA, cutting back into the coronary sinus could add to this further. Once the anatomic landmarks are defined the suturing of the patch is straightforward, taking care of the conduction pathway.

Consent
Informed consent was obtained from patient’s (minor) family members.

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Conflicts of interest
There are no conflicts of interest.

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