

Double orifice mitral valve (DOMV) with atrioventricular canal defect (AVCD) - *A surgical challenge*

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Abstract

Double orifice mitral valve (DOMV) is an unusual finding, most commonly seen with atrioventricular canal defect (AVCD). Due to the varied anatomical types, hemodynamic effects and co existence of other malformations, the surgical management of this anomaly can be challenging. Achieving a compromise between residual regurgitation and new stenosis, should be the aim of surgical intent. We present a case report of DOMV detected incidentally during surgery in a case of AVCD with severe MR.

Text

Introduction

Double orifice mitral valve (DOMV) is a rare cardiac anomaly. It is uncommon to find it as an isolated lesion and is seen associated with atrioventricular septal defects (AVCD) in approximately 5% of cases.¹ Although a number of case reports of DOMV have been reported²⁻⁴, literature highlighting its surgical management is scarce. The challenge lies in, repair of DOMV associated with regurgitation, without producing new stenosis, as inherent nature of this valve predispose it to stenosis. We describe a case report of DOMV associated with AVCD, which was detected incidentally intraoperatively in a 2 year male child with severe mitral regurgitation (MR) and was managed successfully.

Case Report

Informed written consent was taken from the patient for publication. A 2 year male child presented with history of recurrent chest infection and failure to thrive, since 6 months of age. On examination, along with pre cordial bulge, a pansystolic murmur, was heard over left lower sternal border. Echocardiography revealed complete AVCD, moderate tricuspid regurgitation (TR), severe MR and dilation of RA and RV. Since room air saturation was 90% and echo revealed bidirectional flow, cardiac catheterization was done to assess for operability. Baseline Qp / Qs was 2.173 and ratio of pulmonary vascular resistance (PVR) to systemic vascular resistance (SVR) was 0.347. Post oxygen supplementation, the Qp/Qs was 15 and the ratio PVR/SVR dropped to 0.036. Further, ventricular septal defect (VSD) was almost closed; hence patient was labeled as a case of transitional AVCD with severe MR with moderate TR with operable hemodynamics.

Pre-operative on table trans-thoracic echocardiography (TTE) revealed large OP ASD and severe MR with MR jet directed primarily into RA through OP ASD (Figure 1 A, 1 B). In view of these two, less volume was directed towards LV. Patient was operated via standard midline sternotomy with aorto-bicaval cannulation and cardioplegic arrest. Intra-operatively, there was a large ostium primum atrial septal defect (OP-ASD) of size ~ 3×1 cm. No VSD could be appreciated. Left AV valve revealed a double orifice mitral valve (DOMV) (Figure 2 A), with two orifices, a major orifice and a minor orifice, divided by a fibrous bridge. We were unable to categorize the valve leaflets of the major orifice, as per standard classification of left AV valve

.On inspection of the subvalvular apparatus, single papillary muscle was present below both the orifices and almost all the chordae were attached to them , making both orifices prone to stenosis after repair. On saline jet test, minor orifice was competent, but gross MR was seen to be emanating from cleft like area in major orifice .The bridge between the two orifices was left untouched. The cleft like area in the major orifice was partially closed with 6/0prolene, using continuous suture technique. After repair, sizing of the two orifices was done with Hegar's dilator and the combined orifice area of the two orifices corresponded to z value of 0 according to normogram (Figure 2 B). Saline jet test post mitral valve repair revealed no significant MR. OP-ASD was closed with pericardial patch leaving coronary sinus in RA. Post operative, needle estimation revealed, pulmonary arterial pressure (PAP) of 35/23 mm Hg (29) and left atrial pressure (LAP) of 8 mm Hg. Intra operative TEE (Figure 3 A, B) confirmed no residual ASD, mild MR, no mitral stenosis (MS) and mild tricuspid regurgitation (TR). In view of increased volume load delivered to LV now, the morphology of left AV valve was more appreciable. Post-operative course was uneventful.

Discussion

DOMV is a rare congenital cardiac malformation and around 5% of cases of AVCD have associated DOMV with them.¹ The above entity can present itself as a normal functioning valve or less commonly, as mitral regurgitation (MR) or mitral stenosis (MS).⁵

The characteristic feature of DOMV is that, all the chordae of one papillary muscle go to one of the ostia while all of the chordae of the other papillary muscle go to the other ostia.⁶ This predisposes to stenosis if the cleft is extensively sutured, creating a parachute like configuration. The degree of valve narrowing (minimum orifice area), which is acceptable has not been determined, however, orifice of 65 to 70 % is well tolerated.⁷

Our patient had DOMV, AVCD, severe MR with major orifice only slightly larger than accessory orifice. .We opted for sequential approach, starting from the periphery of the major orifice and then moving towards the centre, thereby testing at each step, adequacy of the orifice to avoid stenosis. .We used Hegar's dilators intra-operatively, to objectively assess the diameters of orifices, in order to calculate the area of both orifices for inflow into left ventricle (LV). It is preferable to accept mild regurgitation than to create a new significant stenosis.

As expected, treatment for non regurgitant cleft in DOMV, AVCD is controversial. Some may argue against closing the cleft, to create new stenosis, however, partial closure can prevent late development of MR. Accessory orifices are seldom more than mildly incompetent and hence can be left untouched. Suture or patch closure of regurgitant accessory orifice has been practised, but we believe, ratio of the two orifices must be taken into consideration before this kind of intervention.⁸

Watchful closure of the cleft to avoid new stenosis can be done in DOMV, AVCD, severe MR, with good early outcome, however late development of hemodynamically significant MR is always a possibility.

Author contributions

Dr. Sachin Mahajan - Conception and design.

Dr. Sudhansoo Khanna - Data analysis, Drafting article

Dr. Pankaj Aggarwal – Critical revision of article.

Dr. Vikram Halder - Data collection.

Dr. Nischitha Gowda – Data collection.

References

1. Rosenberg J, Roberts WC. Double-orifice mitral valve Study of the anomaly in two calves and a summary of the literature in humans. Arch Path. 1968 Jul; 86(1):77-80.
2. Buuren FV, Faber L, Bogunovic N. Double orifice mitral valve with a normal function: an echocardiographic and MRI study of a rare finding. Eur Heart J 2011;32:137.

3. Westendorp IC, de Bruin- Bon HA, Hrudova J. Double orifice mitral valve; a coincidental finding. Eur J Echocardiogr 2006;7:463– 4.
4. Congin S, Josa M, Friexa X, Azqueta M, Mestres C, Mulet J. Mitral insufficiency with double- orifice mitral valve in an adult patient. J Thorac Cardiovasc Surg 2007;134:250 –1.
5. Trowitzsch E, Bano-Rodrigo A, Burger BM, Colan SD, Sanders SP. Two-dimensional echocardiographic findings in double orifice mitral valve. J Am Coll Cardiol. 1985 Aug; 6(2):383-7.
6. Van Mierop LHS: Pathology and pathogenesis of endocardial cushion defects. Surgical implications, Second Henry Ford Hospital International Symposium on Cardiac Surgery, Chap 34, JC Davilla, ed., New York, 1977, Appleton-Century Crofts.
7. Lee CN, Danielson GK, Schaff HV, Puga FJ, Mair DD. Surgical treatment of double-orifice mitral valve in atrioventricular canal defects. Experience in 25 patients. J Thorac Cardiovasc Surg.1985;90(5):700-705.
8. Pontailier M, Haidar M, Me´ot M, Moreau de Bellaing A, Gaudin R, Houyel L et al. Double orifice and atrioventricular septal defect: dealing with the zone of apposition. Eur J Cardiothorac Surg 2019; doi:10.1093/ejcts/ezz085.

Figure legends

Figure 1 A: Pre-operative trans-thoracic echocardiography (Apical 4 chamber view) showing large OP-ASD with severe MR.

Figure 1 B: Pre-operative trans-thoracic echocardiography (Apical 4 chamber view) showing tricuspid regurgitation.

Figure 2 A: Intra-operative picture showing double orifice mitral valve with bridging tissue in between two orifices.

Figure 2 B: Intra-operative picture showing the two orifices after the partial closure of cleft in the major orifice.

Figure 3 A: Post-operative TEE (Trans gastric basal short axis view) showing double orifice mitral valve.

Figure 3 B: Post-operative TEE continuous wave doppler (Mid esophageal long axis view) showing gradient of 5/2 mm Hg across mitral valve.





