

Doubly committed subarterial ventricular septal defect with prolapsed right coronary cusp with moderate aortic regurgitation

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Article Info

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Abstract

A 4 year old girl was presented with the respiratory tract infection, breathlessness after taking meal, failure to thrive, abnormal movement of the chest on left side overlying the area of heart and systolic murmur. She developed these symptoms gradually for the last 3.5 years. Echocardiography revealed doubly committed subarterial ventricular septal defect with moderate aortic regurgitation. The size of the ventricular septal defect was 7 x 9 mm at the left ventricular outflow tract. The right coronary cusp of the aortic valve was prolapsed. Left atrium and left ventricle were dilated. The pulmonary artery systolic pressure was 35 mm Hg. The ventricular septal defect was closed with the standard surgical procedure using cardiopulmonary bypass followed by aortotomy and right atriotomy. Immediate post-operative period of this case was uneventful and the patient was discharged on 9th post-operative day. Follow-up echocardiography showed no residual ventricular septal defect or aortic regurgitation and the ventricular function was good.

Introduction

In the western population doubly committed and juxtra-arterial ventricular septal defects are usually found in one twentieth of all ventricular septal defects closed surgically. But in the Eastern population these type of defect is found in one-quarter of all surgically treated ventricular septal defects.¹ In the normal heart, muscular subpulmonary infundibulum lifts the pulmonary trunk away from the base of the heart. In above mentioned type of defect lacking of the infundibular sleeve is found. In that situation fibrous continuity is observed between the aortic and pulmonary valve leaflets. Due to deficit muscular support of right coronary leaflet of the aortic valve, it is prolapsed into the defect, venturi effect also pulls the leaflet towards the flow, both factor results in aortic insufficiency.^{1,2}

Ventricular septal defects in this location are bordered in part by a space which overlies the pulmonary and aortic valve. When viewed from the left ventricular aspect, these defects are in the outflow portion of the ventricular septum, beneath the right coronary cusp.^{1,2}

An important echocardiographic finding is aortic and pulmonary valves lies at the same level in the presence of sub-arterial or doubly committed ventricular septal defects, rather than the pulmonary valve being elevated above

relative to the aortic valve. This provides a diagnostic clue in echocardiography and angiography, along with the absence of outlet or infundibular septum. Echocardiography can also demonstrate associated prolapse of aortic cusp and aortic regurgitation with this type of ventricular septal defects.^{2,4}

Case Report

A 4 year old girl from Comilla was admitted at the Department of Cardiac Surgery, four months back with the past history of respiratory tract infection with high fever at her age of 6 months. She was hospitalized at that time and cured with medical management. On cardiac auscultation her attending physical found murmur and referred her in a cardiac hospital in Dhaka but the parents did not attain. As the time passed her condition deteriorated. She used to become restless after taking meals and failed to grow properly. They noticed abnormal movement of the chest overlying the heart as well as abnormal sound. So, they decided to come in outpatient department of Bangabandhu Sheikh Mujib Medical University as per advice of a cardiologist in her locality. She was diagnosed as a case of ventricular septal defect with aortic regurgitation. Thoracic echocardiography (Figure 1) revealed



doubly committed sub arterialventricular septal defect with aortic regurgitation. Size of the ventricular septal defect was 7 mm x 9 mm at left ventricular outflow tract, with left to right shunt, with peak pressure gradient across the shunt was 75 mm Hg. There was prolapsed right coronary cusp of aortic valve with moderate aortic regurgitation. Left atrium and left ventricle was dilated. Pulmonary artery systolic pressure was 35 mm Hg. After preparation for surgery she underwent intra-cardiac repair operation. Chest was opened with standard median sternotomy. Cardiopulmonary bypass was established with bi-caval cannulation of appropriate size. Heart was arrested by adminis-

tration of antegrade blood cardioplegia with moderate hypothermia. Aortotomy was done by an oblique incision over the aorta away from the orifice of coronary arteries. Right coronary cusp was found prolapsed (Figure 2) and was repaired by Trusler's technique of repair. Aortotomy was closed. Then right atriotomy was done and a moderate size outlet ventricular septal defect was found. Ventricular septal defect was repaired with a Dacron patch. Right atriotomy was closed. De-airation was done before the closure of aortotomy and right atriotomy incision. Patient was weaned from cardiopulmonary bypass uneventfully and chest was closed following standard protocol. Aortic cross clamp time was 54 min and total cardiopulmonary bypass time was 84 min. The post operative period was uneventful. The general condition of the patient improved and her symptoms resolved. The patient was discharged on 9th post-operative day with advice for follow-up.

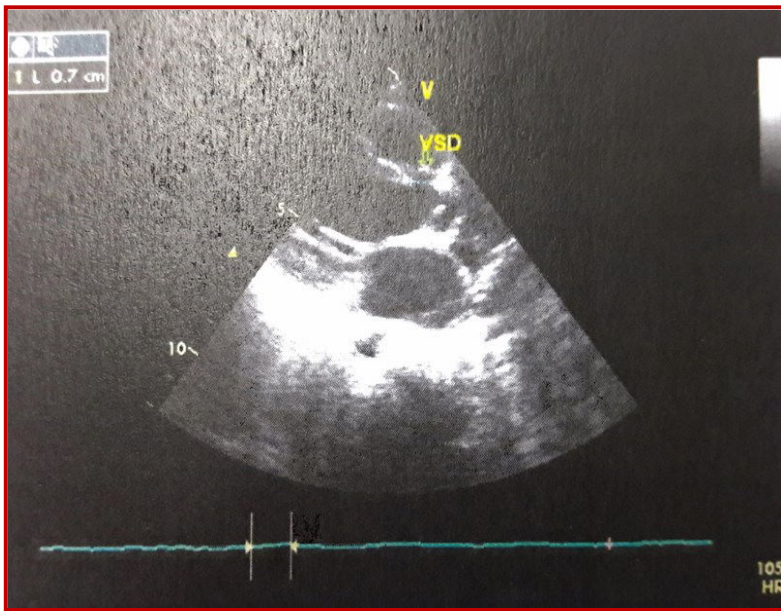


Figure 1: Transthoracic echocardiography showing presence of doubly committed sub arterial ventricular septal defect with moderate aortic regurgitation

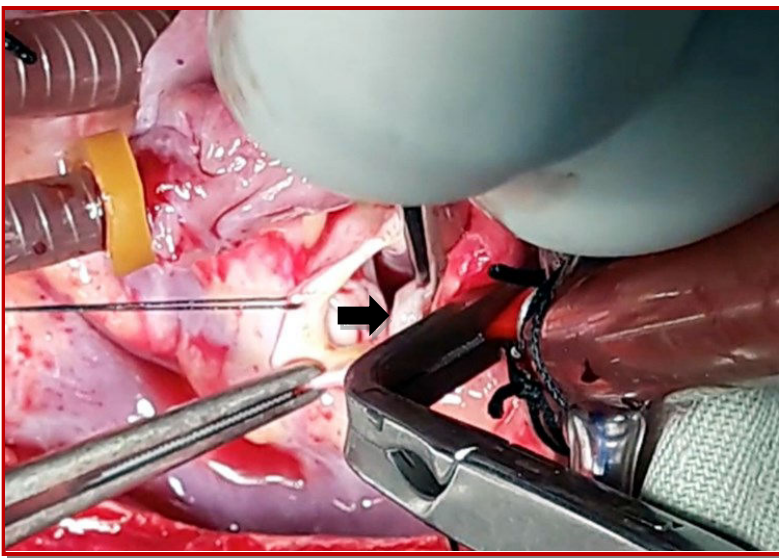


Figure 2: Arrow indicating prolapsed right coronary cusp (RCC) of aortic valve seen through aortotomy

Discussion

This patient was presented with respiratory tract infection, breathlessness after taking meal, failure to thrive, abnormal movement of the chest on left side (area overlying the heart) and systolic murmur. Echocardiography revealed doubly committed sub arterial ventricular septal defect with moderate aortic regurgitation. Size of the ventricular septal defect was 7 mm x 9 mm at left ventricular outflow tract. Right Coronary Cusp of aortic valve was prolapsed. Left atrium and left ventricle was dilated. Pulmonary artery systolic pressure was 35 mm Hg.

Among various subtypes of ventricular septal defect, doubly committed sub-arterial type is important because it has high prevalence in Asian population (30%) as compared to western population (5-10%) and it is associated aortic valve prolapse and aortic regurgitation which is estimated to exceed 40%. The peak age for aortic valve prolapse was around 7 years and that for aortic regurgitation was between 5 to 10 years.⁵

Lun et al. observed that aortic cusp deformities and AR does not develop in sub-arterial type of ventricular septal defects measuring <5 mm in diameter.³ In defects larger than 5 mm, occurrence of the above mentioned complications can be prevented by doing early surgical closure which is supported by other authors also.^{4,6}

The incidence of severity of aortic regurgitation can be reduced by performing surgical closure at proper time and thus need aortic valve surgery, aortic valve replacement, development of pulmonary artery hypertension and its consequences can also be reduced.^{7,8} When surgery is differed in patients with small size or due to other cause serial two

dimensional echocardiography and clinical observation of appearance of aortic regurgitation murmur should be monitored.[1,6-8,10](#)

Prevalence of aortic valve prolapse (herniation) increased with age. Increase in aortic valve herniation is associated with increase in the prevalence of aortic regurgitation. In a study carried out by Schmidt mentioned that patient age at repair is not important. Patient does not develop aortic regurgitation in post-operative period when herniation without regurgitation was present preoperatively. Otherwise, aortic regurgitation is found frequently in post-operative period in those patients having both herniation and regurgitation before surgery. Some authors mentioned that early surgical closure of the defect can prevent the progression of aortic regurgitation.[1,4-6,10-12](#)

We have presented this case because ventricular septal defect associated with prolapsed right coronary cusp of aortic valve and aortic regurgitation is a rare disease. In this case, ventricular septal defect was closed with standard surgical procedure using cardiopulmonary bypass followed by aortotomy and right atriotomy that is mentioned in other study also.[8-10](#) Immediate post-operative outcome of our case was uneventful and the patient was discharged on 9th post-operative day. Follow-up echo showed no residual ventricular septal defect or aortic regurgitation and ventricular function was good.

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