Mitral Valve Replacement with Preservation of Subvalvular Apparatus in a Patient with Familial Dextrocardia and Situs Solitus

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ABSTRACT

Familial dextrocardia with situs solitus is extremely rare entity. Dextrocardia offers a difficult situation to surgeon. A different strategy for cannulation and approach to the left atrium has to be followed. Surgery in such settings has rarely been reported. We present the case of a patient with dextrocardia and situs solitus wherein a left sided approach was adopted for a better exposure of the valve. The patient had a rheumatic regurgitant mitral valve with ruptured chordae to anterior mitral leaflet. Since the valve was severely thickened, it was replaced rather than repaired. The left sided approach provided good exposure of the valve.

Key Words: Mitral valve replacement. Dextrocardia. Situs solitus.

INTRODUCTION

Dextrocardia is a rare cardiac anomaly in which the apex of the heart is pointing towards the right side of the chest rather than the left side.1 There are many types of dextrocardia, mostly associated with other anomalies like reverse position of the abdominal organs. The commonest type is the one where the heart is only a mirror image of its original shape. Dextrocardia offers an unusual situation to cardiac surgeon. A different strategy has to be devised for cannulation and approach to the pathology.

We report a case of a patient with familial dextrocardia and situs solitus where underwent mitral valve replacement for rheumatic heart disease with preservation of subvalvular apparatus.

CASE REPORT

A 33 years old patient presented to our hospital with a history of exertional dyspnoea, fatigue and palpitations. Family history showed that 2 of her 3 siblings had the same anomalous location of the hearts. An initial evaluation with chest X-ray showed typical finding of heart towards the right side.

Transthoracic echo revealed severe mitral regurgitation, ruptured chordae to anterior mitral leaflet in a dextrocardiac heart. The patient had moderate pulmonary hypertension. The leaflets were thickened. Abdominal ultrasound showed normal location of the abdominal viscera. The siblings were investigated and their abdominal viscera was also found normal. A decision was made to replace the valve. A left sided approach was adopted with the surgeon standing on the left side of the patient. A right sided aortic arch was found during the operation. The pulmonary trunk was on the right of the aorta (Figure 1).

![Figure 1: The pulmonary trunk is on the right side of the aorta. RV = Right ventricle; RA = Right atrium; PT = Pulmonary trunk.](image1)

![Figure 2: Mitral valve in situ. The postoperative mitral leaflet oriented to the left side of the patient. AML = Anterior mitral leaflet; PML = Posterior mitral leaflet.](image2)
An ascending aortic and superior vena cava cannulation was done. The left atrium was huge with a size of 60 x 85 mm. A left atriotomy was done from the left side of the patient. The valve was assessed. A change in the position of the mitral leaflets was observed. The anterior mitral leaflet was to the right side while the posterior was oriented to the left side of the patient (Figure 2). The valve was replaced with metallic prosthesis preserving the subvalvular apparatus using a modified Okita procedure of preservation of subvalvular apparatus. The patient had an uneventful postoperative course.

**DISCUSSION**

The incidence of dextrocardia associated with *situs solitus* is 1:30,000 live births. Compared to dextrocardia with *situs inversus*, this anomaly is extremely rare. Cardiac surgery in patients with dextrocardia poses difficult problems owing to the unusual anatomy of the heart. Mitral valve replacement in the settings of dextrocardia with *situs solitus* is extremely rare with only a few cases reported in literature.

Familial dextrocardia is an extremely rare finding and very few cases are reported in the literature. Fishman *et al.* have reported a family with major cardiac anomalies including dextrocardia and ocular anomalies in 3 members. But in this family, the dextrocardia was of the *situs inversus* type. This patient had 3 siblings with dextrocardia and all of them with *situs solitus*.

The mitral valve was approached from the left side of the patient with the incision in the lateral wall of the left atrium. Kikon *et al.* also reported a case of dextrocardia with *situs solitus* with the surgeon approaching from the left side of the patient, just as in this patient. The left sided approach makes it easier to visualize the valve as it will appear from the right side. The left sided approach has been found comfortable by others.

Various surgical techniques have been used to deal with the unusual anatomical disposition of the heart and get good exposure of the valve. In this case, the patient had a moderate pulmonary hypertension. The aorta was difficult to be visualized and consequently cannulation was not easy. But two-stage venous cannula was used for venous drainage, without inferior vena cava cannulation. An inferior vena cava cannulation could have been extremely risky because of the fact that the left ventricular would be lifted up which could have caused its rupture against the valve prosthesis. The incision was made in the left atrium with the surgeon on the left side of the patient. This approach was noted to provide a good exposure and less retraction that was required. A star fish retractor has been used in cases reported previously to lift the heart to the left side and establishing cardiopulmonary bypass.

This report provides an account of a rare case of dextrocardia with *situs solitus* where mitral replacement was performed for severe mitral regurgitation. It can be concluded that the left sided approach provides a good exposure of the valve with little need of extra instrumentation like star fish retractors. The inferior vena cava cannulation should be avoided.

**REFERENCES**