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Double Valve Replacement in Dextrocardia: Unusual Surgery in a Rare Anomaly

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Abstract

Dextro cardia is a rare heart condition of 0.37 to 0.53% 10,000 live births. 45 year old patient had successful double valve replacement (AVR and MVR) with moderate pulmonary hypertention in this anomaly and is extremely rare. The altered surgical anomaly and the necessary changes in surgical approach are described. This procedure is rarely done and is reported very few in the literature to our knowledge.

Keywords: Dextrocardia; DVR; Situs Inversus.

Introduction

Dextrocardia is a rare heart condition characterized by abnormal positioning of the heart in which the tip of the heart (apex) is positioned on the right side of the chest. The position of the heart chambers as well as the visceral organs such as the liver and spleen is reversed in (situs inversus). Most affected individuals can live a normal life without associated symptoms or disability. The prevalence rate of dextrocardia[1,2] is ranged from 0.37-0.53% per 10,000 live birth.

Dextrocardia is believed to be transmitted by autosomal recessive genes [5]. The risk of transmitting the disease to the children of a couple, both of whom are carriers for a recessive disorder, is 25 percent.

Dextrocardia can also be associated with other heart defects like double outlet right ventricle, pulmonary stenosis or atresia, single ventricle, transposition of great arteries and endocardial cushion defects [6].

Our case report is a 48 years old male patient with dextrocardia who was diagnosed to have Mixed lesion of aortic and mitral disease with moderate elevation of pulmonary artery pressure,operated for double valve replacement. We have chosen to report this case because it is the first one in our country and also worldwide only few such cases have been reported.

Material and Methods

A 48 years old man was referred to our centre ten years ago for valvular heart disease with dextrocardia. At that time he had a moderate mitral stenosis and mild aortic regurgitation. He was having mild exertional dyspnea and was in NYHA II. He was on regular follow up and his medications included enalapril, digoxin and Lasix.

Two years ago he had an onset of atrial fibrillation which was satisfactorily well rate controlled with metoprolol and cordarone. His medications were readjusted, Few months back he started feeling some exhaustion after routine efforts and his cardiac echo showed severe mitral stenosis, grade II aortic

regurgitation, dilated LA (56 mm) and dilated LV (LVED 60 mm) EF was 60 %.

In view of his progressive disease and risk of further LV dysfunction, DVR was proposed as his treatment plan. The timing and benefits of the operation were explained to the patient as well as probable risks and complications.

A preoperative angiography showed normal coronaries and confirmed a preserved LV function. An abdominal CT confirmed the type of dextrocardia which was a situs inversus totalis. Midline sternotomy was performed. Surgeon was on the left side of patient and assistant on the right. Dextrocardia anatomy was a mirror image of routine anatomy and all exposure were adequate.

Routine aortic and bicaval cannulations were done and CPB set up after heparinization. Cardioplegic arrest was achieved by direct antegrade blood cardioplegia into the coronary ostia after aortotomy since patient had AR. Topical cold ice cardioplegia was also used and moderate hypothermia applied.

MVR was performed through LA approach, which was easily accessible by the surgeon on the right side of the patient.mitral valve was thick, fibrotic and fused and looked like rheumatic etiology. MVR was done using ATS mechanical mitral valve prosthesis 25mm, intermittent ethibond 2/0.

AVR was performed using a St Jude mechanical aortic valve prosthesis 19mm, aortotomy was closed routinely in 2 layers.

Weaning off bypass was smooth and uneventful with some dobutamine support.

Postoperative recovery was smooth and patient was discharged home on post op day 7. Since then he has been on regular outpatient follow up and doing fine.

Discussion

Dextrocardia may be associated with other cardiac or non cardiac anomalies and therefore it was important in our patient to rule out any such anomalies before surgery. In situs inversus [7], viscera which are normally present on the right side are situated on the left, while viscera which are normally on the left are situated on the right. In such patients, the great vessels as well as the superior and inferior vena cava may show variations which had to be well investigated prior to surgery.

A detailed transthoracic cardiac echo or trans esophageal echo is mandatory for an accurate diagnosis and avoid any unexpected finding during surgery. A CT image is also important in elucidating the type of dextrocardia and confirm any other existing anomaly. The surgeon also had to pay particular attention to the whole anatomy to ensure surgery proceeded in the proper way.

In the simplest type of dextrocardia, the heart is a mirror image of the normal heart and there are no other problems. The organs of the abdomen and the lungs will also be arranged in a mirror image. In more complex forms the abdominal and chest organs may be abnormal and may not work correctly.

A very serious syndrome that appears with dextrocardia is called heterotaxy[3,4]. In this condition, many of the organs are not in their usual places and may not work properly. For example, the spleen may be completely missing or exist as several small spleens and do not work properly. Heterotaxy can also be associated with abnormal gall bladder system, anomalies of the intestines as well as anomalies of the blood vessels. Possible risk factors for dextrocardia include a family history of the condition.

The incidence of CAD in population with dextrocardia is similar to that of the general population [8] and in our case a preoperative angiography confirmed normal coronaries.

The timing of surgery was another aspect of consideration for the proper treatment of our patient. He has been on regular follow up at our centre for the past ten years. It was also important for the surgeon to decide on which side he would be during operation. Our patient was a typical case of dextrocardia with total situs inversus and all the visceral organs were mirror images of one another.

Being on the left side of the patient was more appropriate for the surgeon to have a more natural view of the anatomy of the heart. Exposure And of the cannulation sites were also easier to approach. St Rammos et al [9] established CPB by cannulating the aorta and the left common femoral vein. In our case, CPB was set up routinely after cannulating aorta, superior vena cava and inferior vena cava. Yokoyama et al [10] reported a case of double valve replacement in a patient with dextrocardia and situs inversus in which they stood on the left side of the patient.

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