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MITRAL VALVE REPLACEMENT IN DEXTROCARDIA WITH SITUS INVERSUS TOTALIS: A CASE REPORT

Internal Medicine

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ABSTRACT

situs inversus with dextrocardia is a rare congenital anomaly. This is a reverse isomeric form of the thoracic and abdominal viscera or the complete mirror image. Acquired mitral valve disease that requires surgery in a patient with dextrocardia and situs inversus is an exceptional finding. The transseptal approach for mitral valve surgery in dextrocardia represents a technical challenge owing to its anatomic particulars. Here, biatrial cannulation was performed with the surgeon standing on the patient's right side, and mitral valve replacement using a transseptal approach was performed with the surgeon standing on the patient's left side. In this case report, we emphasize the rarity of mitral valve disease in a patient with dextrocardia and the inherent potential difficulty that appears in this particular anatomic condition.

KEYWORDS

dextrocardia, situs inversus totalis, mitral valve replacement.

INTRODUCTION:

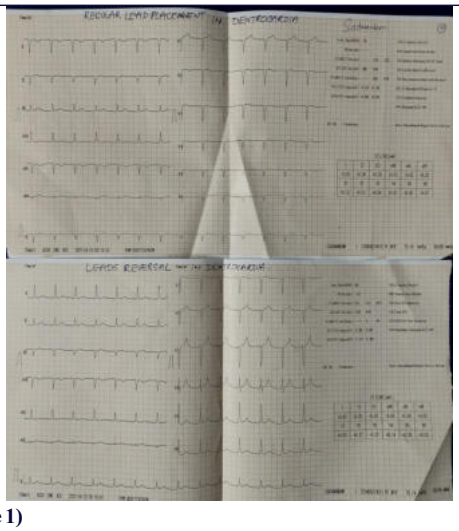
Dextrocardia is a rare congenital cardiac anomaly in which the heart is located in the middle of the mediastinum, with its base to the apex directed to the right and caudal. In different studies, the prevalence rate of dextrocardia ranges from 0.37% to 0.53% per 10000 live births. Situs inversus totalis associated with dextrocardia has a rare incidence of 1 in 10000 adult population (Garg 2009). The concept of situs refers to the configuration of asymmetric organs in the human body. Anatomically, 3 types of situs have been described: situs solitus, which is the normal form of the placement of the organ in the human body; situs inversus, which is a mirror image of the normal placement of organs, anatomy, and morphology of the abdominal and thoracic organ and vessel systems are conserved, the left atrium is located to the right, the left lung has 3 lobes and the right lung has 2 lobes, the stomach and the spleen are positioned on the right, and the larger lobe of the liver is on the left side of the body. The causes of dextrocardia have not yet been identified, but it was observed that a quarter of patients with the pathological condition have associated primary ciliary dyskinesia (Kartagener syndrome) with autosomal recessive inheritance (zariwala 2013;moldjian 2007) dextrocardia is not directly associated with race, ethnicity, or sex.

Dextrocardia in adults is usually diagnosed incidentally during routine or radiological examination for some other purposes.

Here in, we present a case of situs inversus totalis and dextrocardia with severe mitral regurgitation, who underwent mitral valve replacement with 29mm no TTK Chitra tilting disc mitral valve in our institute.

Case Report:

A 63-year-old male patient with hypertension presents to our institution with palpitation and dyspnea for 1 month. He was in the class 3 New York Heart Association. Electrocardiography (ECG) showed a normal sinus rhythm (Figure 1). Chest x-ray showed that the heart was directed to the right with mild cardiomegaly. Carotid Doppler ultrasonography and blood test results were normal. The patient was diagnosed with mitral valve prolapse, severe mitral regurgitation with mild stenosis by transthoracic and transesophageal echocardiography. He had a left ventricular end-systolic diameter of 3.6 cm and an end-diastolic diameter of 4.8 cm and an ejection fraction of 45%. He had a left atrial anteroposterior diameter of 5.5 cm. The coronary arteries were normal in the preoperative coronary angiography. (Figure 2). During the preoperative period the contrast chest and abdomen CT (Figure 3) showed both dextrocardia and situs inversus totalis; with normal continuity of the inferior vena cava.



(Figure 1)

(Figure 2)

(Figure 3)

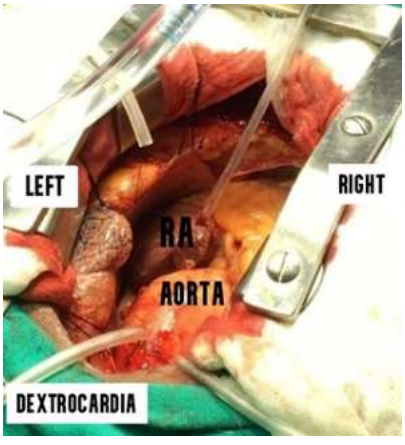
Surgical Technique:

After sufficient anesthesia, following midline sternotomy and

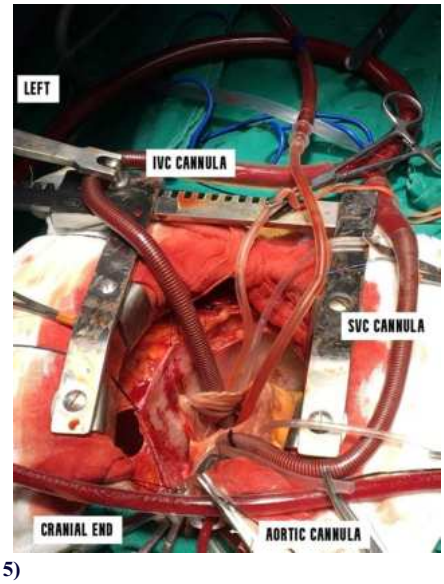
pericardiotomy, the superior vena cava and inferior vena cava were widely separated from the pericardium for cannulation and decannulation without lifting the heart (Figure 4). Arterial cannulation was performed on ascending aorta and venous cannulation was done through bicaval canulation of the superior and inferior vena cava (Figure 5). Moderate hypothermia (28 degrees Celsius) was applied. The surgeon switched his position from the right to the left side of the patient after establishing of cardiopulmonary bypass. After aortic cross-clamping, antegrade cold (6 to 8 degree C) delnido cardioplegia was administered while venting was performed through the left atrium.

The incision on the right atrium was parallel to the atrioventricular groove. following the right atriotomy, a septal incision was performed in the center of the fossa ovalis. Silk retracting sutures were placed on the septal edge of the septum and RA valves. Despite these maneuvers, the mitral valve was exposed with great difficulty. Intraoperative findings revealed that the mitral valve was affected by myxomatous changes and PML prolapsed, repair of the mitral valve was not feasible. The anterior leaflet was excised while the posterior leaflet was preserved. Mitral valve replacement by 29mm. TTK Chitra tilting disc mechanical prosthesis was done using pledgetted 2-0 polyester suture in an intermittent everting mattress fashion (Figure 6). The interatrial septum closed with 4-0 prolene, and RA is closed with 5-0 prolene suture. After de-airing, the aortic cross-clamp was removed and the heart started beating spontaneously. He was gradually weaned off from CPB, on moderate ionotropic support. The total CPB time was 180 min, and the aortic cross-clamp time was 120 min.

The patient's postoperative course was uneventful, and he was discharged from the hospital on the 5th post-operative day, post operative chest x-ray was normal (Figure 7). Transthoracic postoperative echocardiography revealed a mechanical prosthesis with a mean gradient of 3mm across the prosthesis and normal left and right ventricular function.



(Figure 4)



(Figure 5)

(Figure 6)

(Figure 7)

DISCUSSION:

Dextrocardia is a rare congenital anomaly and was first described by Hieronymus Fabricius in 1606. Although dextrocardia can be associated with situs solitus, situs inversus, or situs ambiguous; situs solitus is the most common form. Situs inversus totalis, as in our case, is a reverse isomeric form of the thoracic and abdominal viscera or a complete mirror image. Uchimiro et al (2002) published the case of a patient with an interrupted inferior vena cava in the intrahepatic segment. These abnormalities may cause problems in the inferior venous cannulation. In a similar case, a supplementary venous cannula was inserted into the hepatic vein from the right atrium (koga 2019). Therefore, abnormalities in the inferior vena cava must definitely be examined by computed tomography or magnetic resonance imaging.

In our case, the preoperative CT scan showed no abnormalities in the position of the inferior and superior cava at the level of the right atrium; and the dimensions of the right ventricle were almost normal. Established cardiopulmonary bypass by cannulating the aorta and the superior and inferior vena cava separately without using any positioner to lift the heart, since we performed cannulation by widely separating the superior vena cava and the inferior vena cava from the pericardium. This may be a technical trick for safe bical cannulation in dextrocardiac patients.

Due to rare condition and abnormality in the heart position, the surgeon encounters many challenges in such patients, namely dexterity, the position to operate from, establishing CPB, approach to the mitral valve, etc. the importance of understanding surgical anatomy in such a case make strategic planning of surgery and avoidance of procedure-related complications and optimizing surgery output.

We approached the mitral valve via transseptal approach; in general, in other cases, the mitral valve has been accessed through the left atrium

(75%) (Berhane 2016). We believe that our approach by transseptal mitral valve replacement with bicaval cannulation strategy is a sure and reproducible technique that can also be used in patients with associated tricuspid valve disease. Thorough preoperative study and understanding of the anatomy of patients with dextrocardia associated with situs inversusis the key to successful mitral valve surgery.

Our case report is also unique as there are very limited case reports of mitral valve replacement via transseptal approach for severe MR in dextrocardia with situs inversus totalis.

CONCLUSIONS:

We suggest standing on the left side of the patient and to separate the superior and inferior vena cava widely from the pericardium for better exposure to cannulation and decannulation without lifting the heart, which can prevent the danger of left ventricular rupture.

A transseptal approach using an incision at the level of fossa ovalis extended in a cephalad direction toward the left atrial roof direction in a patient with dextrocardia and situs inversus totalis has to be an excellent approach that ensures wide exposure of the mitral valve and no morbidity in the case of the mitral valve replacement.

Declaration Of Conflicting Interest:

The authors declared no conflict of interest with respect to the authorship and/or publication of this article.

Consent:

Informed consent was obtained from the patient to publish this case report for academic interest.

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