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An experience of successful triple valve surgery in mirror image dextrocardia with situs inversus totalis

Situs inversuslu ayna hayali dekstrokardi olgusunda başarılı üç kapak cerrahisi deneyimi

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Summary

Cardiac surgery in patients with a positional anomaly of the heart is technically challenging, and very few reports exist of such surgery in patients with dextrocardia. There are three types of situs: Situs solitus (normal), situs inversus (mirror image of normal), and situs ambiguous. Cardiac surgery for acquired valvular diseases in patients with mirror image dextrocardia and situs inversus is extremely rare. In this article, we report a surgical case of mitral and aortic valve replacement and tricuspid annuloplasty in a patient with mirror image dextrocardia and situs inversus.

Key Words: Dextrocardia, heart valve disease, situs inversus.

Özet

Kalbin pozisyonel anomalisi olan hastalarda kardiyak cerrahi teknik olarak zor olup dekstrokardili hastalarda cerrahi ile ilgili çok az sayıda yazı bulunmaktadır. Üç tip situs mevcuttur; situs solitus, situs inversus (ayna hayali) ve situs ambigus. Ayna görüntüsü situs inversuslu ayna hayali dekstrokardili hastalarda sonradan gelişen kapak hastalıklarında kalp cerrahisi son derece nadirdir. Bu yazıda, aort ve mitral kapak replasmanı ve triküspit annuloplasti cerrahisi uygulanan situs inversuslu ayna hayali dekstrokardili olgu sunuldu.

Anahtar Sözcükler: *Dekstrokardi, kalp kapak hastalığı, situs inversus.*

Introduction

Dextrocardia is a cardiac positional anomaly in which the heart is located in the right hemithorax with its base-to-apex axis directed to the right and caudad. The malposition is intrinsic to the heart and not caused by extracardiac abnormalities. There are three types of situs: situs solitus (normal), situs inversus (mirror image of normal), and situs ambiguous. Situs applies to the pattern of the viscera as a whole and to each asymmetric viscus itself, such as the lung, liver, spleen, and gastrointestinal tract. Situs also applies to the heart as a whole and to each of the cardiac chambers because each is asymmetric (1).

Dextrocardia with situs inversus, L-loop ventricles, and inverted great arteries results from situs inversus with a concordant L-bulboventricular loop. This is the mirror image of normal and has been called “mirror-image dextrocardia” (1).

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This is the most common type of dextrocardia in the general population (present in one or two in 20,000). The incidence of congenital heart disease is low, ranging from 2% to 5% (2). Dextrocardia with situs solitus, D-loop ventricles, and normally related great arteries results from failure of the final leftward shift of the ventricles during embryologic development. This has been termed “dextroversion” because the heart appears to be rotated into the right hemithorax relative to its normal position. Dextroversion is the second most common type of dextrocardia (3).

Valve surgery for acquired valvular lesions in dextrocardia with situs inversus is also rare. However, there is no study in the literature concerning triple valve surgery and dextrocardia with situs inversus totalis. In this case study, we report on a patient with dextrocardia with situs inversus, illustrating the anatomic issues and operative considerations particular to aortic, mitral and tricuspid valve surgery in patients with this condition.

Case Report

A 35 year old pregnant woman was followed medically with the diagnosis of rheumatic valvular (mitral and aortic valves) heart disease with dextrocardia and situs

inversus totalis. At the 28 weeks gestation she was admitted to the cardiology and obstetrics-gynecology department of our hospital with complaints of palpitation and progressive exertional dyspnea. With consent, she was advised to have an immediate birth. The patient subsequently underwent an urgent Cesarean section under general anesthesia and delivered a healthy female infant. She was referred to the cardiovascular department with a diagnosis of aortic and mitral valve stenosis and tricuspid valve insufficiency and compensated heart failure postnatal. The patient was taken to the intensive care unit (ICU). Chest radiograph revealed dextrocardia and discordant location of cardiac apex relative to stomach and liver shadow (Figure-1) and abdominal ultrasound examination confirmed the presence of situs inversus totalis. Findings showed a complete mirror image presentation of the cardiothoracic and visceral organs. Electrocardiogram (ECG) showed normal sinus rhythm (Figure-2).

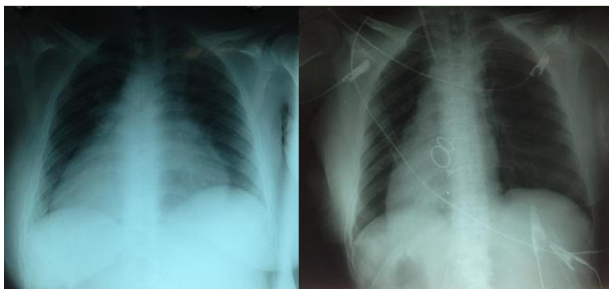

Figure-1. Preoperative and postoperative Chest X-Rays. The cardiac silhouette and gastric air bubble were evident on the right side and liver shadow on the left, indicating situs inversus.

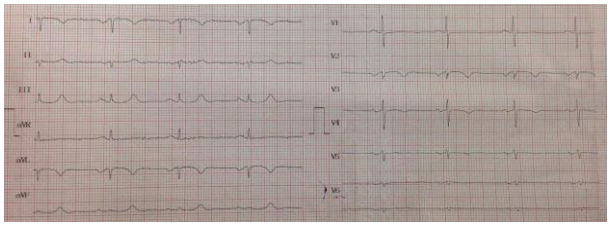

Figure-2. ECG findings. ECG showed a negative P, T and QRS waves in the I and aVL, positive QRS complexes (with upright P and T waves) in aVR. Absent R-wave progression in the chest leads (dominant S waves throughout).

At the transthoracic echocardiography, there was pericardial effusion, pleural fluid, pulmonary hypertension (110 mmHg), severe mitral stenosis (valve area <1 cm2), 3^{o} tricuspid insufficiency, severe aortic stenosis with 2-3^{o} aortic failure and mirror image dextrocardia. New York Heart Association (NYHA) functional class and left ventricular ejection fraction were 3 and 50%, respectively. She was put on medical therapy for prevention of heart failure.

Cardiac surgery was planned five days after the patient's admission to ICU. Mirror-image dextrocardia was seen after median sternotomy (Figure-3) and standard aortic-bicaval cannulation was performed. Cardiopulmonary bypass (CPB) under moderate hypothermia was established. Myocardial management was provided by antegrade intermittent cold and terminal warm blood cardioplegia. A left-sided left atriotomy provided excellent exposure of the mitral valve. After surgical examination, her mitral valve was replaced with a 27 mm St. Jude mechanical valve; and her aortic valve was replaced with a 19 mm St. Jude mechanical valve and tricuspid Kay annuloplasty. She was then weaned from CPB successfully and taken to the ICU. Aortic cross-clamp time was 112 minutes and total cardiopulmonary bypass time was 133 minutes. The patient's post-operative course was uneventful, and she was discharged on the 6th postoperative day in good condition and in sinus rhythm. The patient was followed up for four months with no complaints.

Postoperative transthoracic echocardiograms within the first postoperative week showed normofunctioning aortic and mitral mechanical valves, 1º tricuspid valve insufficiency and systolic pulmonary artery pressure was 30 mmHg.

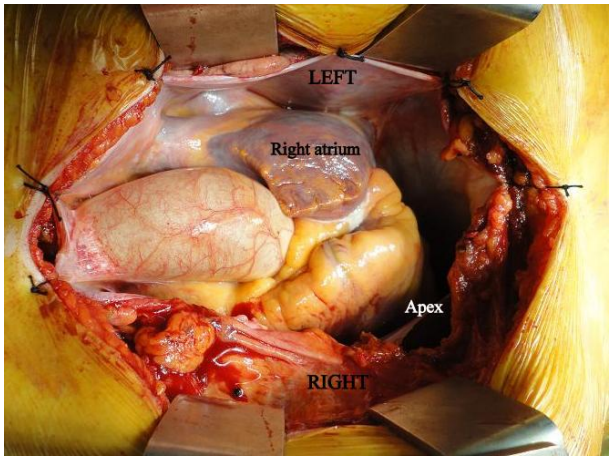

Figure-3. Mirror image of normal.

Discussion

Dextrocardia is a rare abnormality of the heart position (4) Most cases with situs solitus are associated with other cardiac or noncardiac malformations. However, patients with situs inversus totalis (as in our patient) rarely have other associated malformations (4,5). One of the earliest observations of abnormal location of the internal organs is said to have been made by Aristotle (6). Dextrocardia should alert clinicians and surgeons to the possibility of associated cardiac malformations, and a well-established description of the whole situs should

be known prior to surgery. Echocardiography can easily be used to confirm the presence or absence of valvular disease and other cardiac malformations (7). However, there are only a few case reports in the literature on double valve replacement in isolated dextrocardia (8, 9). The case is described of a patient with dextrocardia with situs inversus, illustrating the anatomic issues and operative considerations particular to aortic and mitral valve surgery in patients with this condition. To our knowledge, this is the first case of a triple valve surgery in a patient with dextrocardia with situs inversus totalis. In patients with dextrocardia requiring cardiac valve surgery, it is important to consider the appropriate surgical strategy. Approaching the mitral valve through a left sided left atrial incision seems to provide excellent exposure for mitral valve replacement. In patients with situs anomalies, it is imperative that the cardiovascular surgeon be cognizant of the anomalous anatomy to assure safety and avoid complications.

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