Case report

Redo aortic valve surgery in a case of dextrocardia with situs inversus totalis

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ABSTRACT

Introduction: Dextrocardia is a rare congenital cardiac anomaly where the base-apex axis of the heart is directed to the right side. It may be associated with situs solitus, situs inversus, or situs ambiguous. Such cases are technically challenging when heart surgery is needed.

Presentation of case: We presented a case of a patient with dextrocardia and situs inversus totalis, who had severe aortic bioprosthetic valve degeneration needing a redo aortic valve replacement. Our operative strategy was to change the position of the main surgeon to the left side of the patient to perform most surgical steps.

Discussion: A meticulous pre-operative surgical plan involving the whole team was very important for a smooth intra-operative course and a favourable outcome. Regarding the position of the main surgeon standing on the left side of the patient, we believed this surgical team arrangement added much to the technical ease for the procedure, since the anatomy was opposite to what our minds are used to. Also, computed tomography scan played a crucial role for a proper preoperative anatomical evaluation and surgical planning.

Conclusion: Our approach with the surgeon on the left side provided excellent exposure for redo aortic valve replacement in dextrocardia with situs inversus totalis. This surgical management played an important role for the good result in this technically challenging scenario.

1. Introduction

Dextrocardia is a cardiac abnormality in which the main cardiac axis is directed to the right and caudally. It may be associated with situs solitus, situs inversus, or situs ambiguous. Dextrocardia with situs inversus totalis means the heart and all other visceral organs are the mirror image of normal position. The incidence of dextrocardia with situs inversus totalis is 1/10,000 to 50,000 births [1].

The surgical management of such cases require a good operative planning involving the surgical team arrangement to overcome the technical challenges and achieve an optimal outcome.

Here we report a case of a patient with dextrocardia and situs inversus totalis, who had severe aortic bioprosthetic valve degeneration needing a redo aortic valve replacement. This case is reported in line with SCARE criteria [2].

2. Presentation of case

A 67-year-old woman presented to the emergency room with progressive dyspnea on exertion for 5 months with worsening of symptoms in the last 3 days, with the onset of orthopnea, paroxysmal nocturnal dyspnea and lower limb edema. She had a history of dextrocardia with situs inversus totalis and aortic valve replacement surgery, ten years previously. Her vital parameters were normal, with bibasal crackling sounds in the lungs and a grade 4 systo-diastolic murmur in the right paraesternal area at the 2nd intercostal space.

The chest radiography showed a right-sided cardiac shadow with mild cardiomegaly and signs of pulmonary congestion (Fig. 1A). Transsthoracic ecocardiography revealed dextrocardia, situs inversus, aortic bioprosthetic valve dysfunction due to severe regurgitation and moderate stenosis (mean pressure gradient of 39 mmHg), and a left ventricular ejection fraction of 55%. Computed tomography (CT) demonstrated situs inversus totalis with dextrocardia and no structures

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cases are associated with other congenital cardiac anomalies [3].

... ascending aorta, the superior vena cava, and the inferior vena cava.

In dextrocardia with situs inversus totalis (all visceral organs were mirror images of the normal) and pre-inversus totalis. In dextrocardia with situs inversus, around 10 % of the organs also get mirrored, then it is called dextrocardia with situs inversus totalis. In dextrocardia with situs inversus, around 10 % of the cases are associated with other congenital cardiac anomalies [3].

Our patient had a typical case of dextrocardia with situs inversus totalis (all visceral organs were mirror images of the normal) and presented an aortic biological prosthesis dysfunction requiring a redo aortic valve replacement. The anatomy of the patient is always challenging in the case with dextrocardia and decision on the ideal positioning of the main surgeon is crucial.

Some approaches have been described about patients with dextrocardia with situs inversus totalis for valve replacement but still redo cardiac surgery is rare. Haldar et al. [4] reported a case of aortic and mitral valve replacement in a patient with dextrocardia and situs inversus totalis in which they stood on the left side of the patient. Sahin and colleagues [5] described a transeptal approach for mitral valve replacement. Saad et al. [6] addressed the position of the surgeon in dextrocardia and situs inversus. Similar to our case, Altarabsheh et al. [7] reported a left side approach for aortic valve replacement in a patient with dextrocardia and situs inversus, proposing this operative setting for patients with such unusual anatomy.

A meticulous pre-operative surgical plan involving the whole team was very important for a smooth intra-operative course and a favourable outcome. Regarding intraoperative strategy about the position of surgeon for cannulation and aortic valve approach, we performed bicaval cannulation, prosthesis removing and new aortic bioprosthesis insertion with the main surgeon standing on the left side of the patient. We believed this surgical team arrangement added much to the technical ease for the surgery, since the anatomy was opposite to what our minds believed this surgical team arrangement added much to the technical perspective that left-side approach presents.

In addition, since it was a redo cardiac surgery with an uncommon anatomy, CT scan played a crucial role for a proper preoperative anatomical evaluation of all the great vessels, the heart, possible associated anomalies and to plan our cannulation strategy. Also, CT scan permitted to minimize risk of re-entry injury because it could identify potential adherence of mediastinal structures to the undersurface of the sternum.

3. Discussion

Dextrocardia is a rare congenital cardiac anomaly where the base-apex axis of the heart is directed to the right side. The atrial situs can be situs solitus, situs inversus, or situs ambiguous of which situs inversus (mirror image dextrocardia) is more common (40 %). If all the visceral organs also get mirrored, then it is called dextrocardia with situs inversus totalis. In dextrocardia with situs inversus, around 10 % of the cases are associated with other congenital cardiac anomalies [3].

Our patient had a typical case of dextrocardia with situs inversus totalis (all visceral organs were mirror images of the normal) and presented an aortic biological prosthesis dysfunction requiring a redo aortic valve replacement. The anatomy of the patient is always challenging in the case with dextrocardia and decision on the ideal positioning of the main surgeon is crucial.

Fig. 1. a Chest radiography demonstrating dextrocardia with dilated heart due to left ventricle enlargement and signs of pulmonary congestion. b 3D reconstruction of computed tomography of the chest demonstrating the heart and great vessels exactly a mirror of norm. PA: pulmonary artery; RA: right atrium; RV: right ventricle, SVC: superior vena cava.
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Ethical approval

Ethical approval has been exempted by our institution as this publication is a case report, provided that the patient gave her written consent both for operation and the publication of this case.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor

Fabrício José Dinato

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Pablo Maria Alberto Pomerantzef: critical revision
Fabio Biscegli Jatene: critical revision
All authors read and approved the final manuscript.

Fig. 2. Intra-operative view of the patient showing the aorta and the right atrium (RA) on the left side, and the pulmonary artery (PA) and the left ventricle (LV) on the right side. RV: right ventricle.
Declaration of competing interest

No conflicts of interest.

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Fig. 3. Intra-operative view after prosthesis removal and placing interrupted polyester mattress sutures.