Case Reports

Coronary Artery Bypass Grafting in a Patient With Multivessel Disease and Dextrocardia With Situs Inversus Totalis

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Abstract

Dextrocardia with situs inversus totalis is a rare hereditary condition characterized by reversed orientation of the major thoracic and abdominal organs. Though dextrocardia itself is not believed to increase the risk of coronary artery disease, the workup and surgical management of patients with this condition may be technically challenging to heart team clinicians. This report describes the case management of a high-risk 56-year-old man with dextrocardia who presented with multivessel coronary artery disease.

Keywords: Coronary artery bypass; dextrocardia; situs inversus

Case Report

Presentation and Physical Examination

A 56-year-old man who is a current smoker with a 45 pack-year smoking history presented for evaluation of anginal symptoms, including exertional dyspnea and leg claudication. The patient also endorsed a strong family history of premature coronary artery disease (CAD). He underwent a full workup, including coronary computed tomographic angiography, which showed a calcium score of 1,437 with severe multivessel disease, including the proximal left anterior descending coronary artery (LAD). He was found to have dextrocardia, a finding which he confirmed was diagnosed during his childhood but had never been followed up on because of a lack of symptoms (Fig. 1 and Fig. 2). Electrocardiography showed normal sinus rhythm with lead reversal (Fig. 3). The patient underwent left heart catheterization via the right radial artery and was found to have multivessel CAD involving the distal right coronary artery (80%), proximal and mid-LAD (80%), and the ramus intermedius (70%), with anatomy unsuitable for percutaneous coronary intervention.

Medical History

The patient had a medical history of chronic obstructive pulmonary disease, hypertension, poorly controlled type 2 diabetes (glycated hemoglobin >0.1 of total hemoglobin [>10%]), and morbid obesity.

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Fig. 1 Preoperative chest x-ray shows visible dextrocardia.

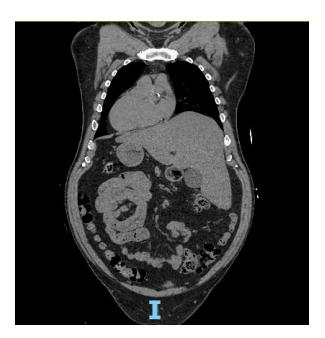


Fig. 2 Preoperative standing computed tomogram of the chest, abdomen, and pelvis without intravenous contrast shows situs inversus totalis.

Differential Diagnosis

A multidisciplinary heart team discussion was initiated, with the interventional cardiology service deeming percutaneous coronary intervention to be complex given the patient's diffuse CAD with dextrocardia. Considering his younger age and the presence of multivessel CAD, including the proximal LAD, and diabetes, the team believed that the best intervention would be coro-

Key Points

- Dextrocardia with situs inversus totalis may complicate the workup and adequate management of CAD in patients with these conditions, requiring multidisciplinary collaboration to evaluate the patient's amenability to percutaneous or surgical revascularization.
- Technical considerations, including careful conduit selection and harvest, operator positioning, and the optimization of pericardial windows for graft insertion, are paramount.
- For revascularization of the LAD, operators may consider the use of the RIMA to avoid excessive manipulation and tension on traditional LIMA grafts..

Abbreviations and Acronyms

CABG	coronary artery bypass grafting
CAD	coronary artery disease
LAD	left anterior descending coronary artery
LIMA	left internal mammary artery
RIMA	right internal mammary artery

nary artery bypass grafting (CABG). After an extensive risk-benefit evaluation and pursuit of a second opinion, the patient and his family consented to surgery, and the patient began a complete preoperative evaluation. Transthoracic echocardiography showed a preserved ejection fraction with mild aortic valve sclerosis. A computed tomogram of the chest, abdomen, and pelvis was notable for situs inversus totalis, with extensive atherosclerotic vascular calcifications within the aorta and coronary arteries. Carotid duplex showed mild stenosis (<40%) bilaterally. His pulmonary function test showed moderate chronic obstructive pulmonary disease. He was estimated to have a class IV risk according to the Revised Cardiac Risk Index, with a 15% 30-day risk of death, myocardial infarction, or cardiac arrest.

Technique

The patient underwent multivessel CABG through a midline sternotomy with harvest of the right internal mammary artery (RIMA) and open saphenous vein harvest to ensure the highest-quality conduits for his non-LAD lesions. Transesophageal echocardiography was used for intraoperative monitoring. The patient was cannulated through the distal ascending aorta, which was on the left side, adjacent to the left pulmonary artery, with venous cannulation through the right atrial appendage, which was also on the left side (Fig. 4). Cardioplegia was given through the antegrade cardioplegia catheter for the induction and subsequent

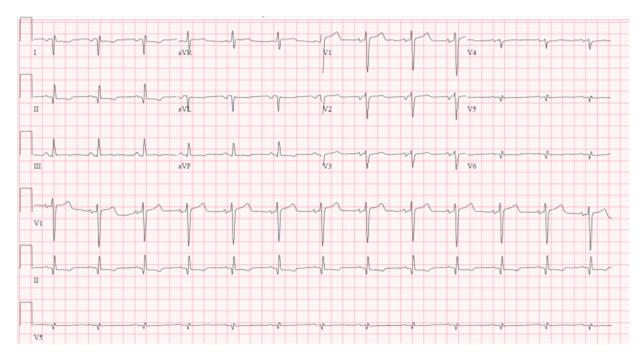


Fig. 3 The patient's electrocardiogram shows normal sinus rhythm with lead reversal.

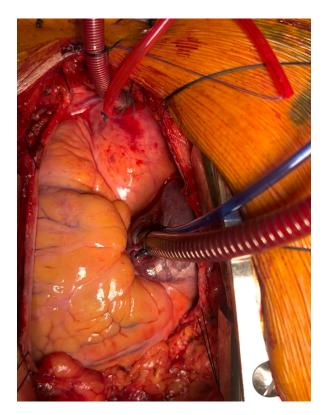


Fig. 4 An intraoperative photograph shows a still frame of dextrocardia with visible right atrial venous cannulation on the left side of torso and adjacent distal ascending aortic cannulation. The supplemental motion image shows the beating heart with bypass catheters.

Supplemental motion image is available for Figure 4.

doses and through the vein bypass grafts after they were constructed. A retrograde cardioplegia catheter could not be used as the coronary sinus was too small on echocardiography to deploy it safely.

First, the right posterior descending artery was bypassed using a reversed saphenous vein graft beyond the acute margin of the heart. This was completed in standard fashion from the right side of the patient. The surgeon then walked over to the left side of the patient to complete the final 2 bypasses: the reversed saphenous vein graft to the ramus intermedius followed by the RIMA to LAD anastomosis (Fig. 5). Gaining an adequate field exposure was challenging given the patient's severe emphysema, morbid obesity, and thick hypertrophied heart. All anastomoses were completed with running 8-0 polypropylene sutures. The patency of each of the grafts was also confirmed with a 1.5-mm probe before tying down the anastomoses. The ramus intermedius graft to the ascending aorta was completed while the surgeon was still on the left side of the patient, and the posterior descending artery was anastomosed in standard fashion from the right side.

Outcome

The patient was successfully weaned from cardiopulmonary bypass and extubated a few hours postoperatively. He was discharged home on post-CABG day 5 without complications.

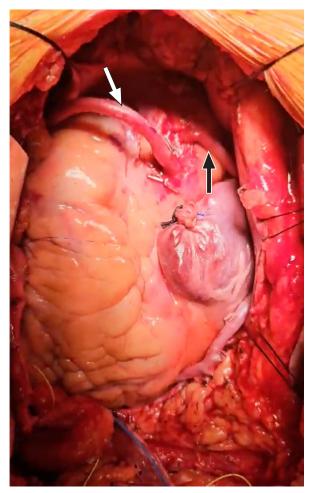


Fig. 5 An intraoperative photograph shows the heart with visible saphenous vein grafts (arrows).

Latest Follow-Up

The patient was seen in the clinic 1 week and 1 month after his operation and went on to make an uneventful and complete recovery.

Discussion

Dextrocardia with situs inversus totalis is a rare congenital condition whereby the orientation of major thoracic and abdominal organs is reversed. Though the condition is not believed to independently increase cardiovascular morbidity, the workup and management of patients with dextrocardia with situs inversus totalis may be challenging, even to experienced heart teams, as operators must adapt their diagnostic, anesthetic, and interventional approaches to accommodate the inversed laterality. Literature on the management of complex CAD in patients with situs inversus totalis is relatively limited, with only 28 case reports describ-

ing 32 patients published up to 2010.2 There are some data to suggest that dextrocardia predisposes patients to further cardiovascular malformations, such as discordancy in atrioventricular connections and decreased pulmonary blood flow.³ Though they are commonly seen in patients with primary ciliary dyskinesia, isolated autosomal recessive mutations to transmembrane and cilia-associated proteins have also been linked to organ laterality defects.4 For adults, the overall prognosis of dextrocardia with situs inversus totalis is good in the absence of other major abnormalities, and patients are expected to have normal life expectancies. The prognosis of dextrocardia in pediatric patients, particularly in the absence of associated situs inversus totalis, is conversely more morbid, with mortality rates varying depending on the severity of cardiac and noncardiac malformations.5

The interpretation of electrocardiographic, echocardiographic, and coronary catheterization images may be particularly challenging. Nakasone and colleagues⁶ described how they successfully modified their anesthesia and imaging setup to mitigate the challenges of perioperative monitoring in patients with dextrocardia. Electrocardiography leads in the operating room should be applied in reverse as standard lead application may misleadingly show a positive aVR lead deflection and right-axis deviation. For continuous hemodynamic monitoring, operators should deploy the Swan-Ganz catheter via the left internal jugular vein to access the right atrium and confirm adequate "mirrored" positioning via radiography. The transesophageal echocardiography probe angle must be modified by 180 degrees for proper windows and views. For instance, midesophageal 4-chamber and transgastric short-axis views would be taken at 180 degrees instead of 0 degrees, with the left ventricle on the left side of the screen, while the aortic valve short-axis view would be taken at 140 to 150 degrees instead of 30 to 40 degrees.⁶ Inverse thoracic anatomy also necessitates modifications in operative planning, including incision access, ambidexterity, and technical nuances, such as landmark orientation and operator table positioning.7

A few centers have previously described the surgical revascularization of patients with dextrocardia. Ravikumar and colleagues⁸ reported the management of a 59-year-old man with dextrocardia and normal global cardiac function. Their patient's condition was managed electively using an arterial graft to the LAD and venous grafts to the D1 artery and ramus intermedius in a manner similar to the one used in the current case.

The primary surgeon operated from the right side of the patient for the majority of the revascularization. A 2010 case report by Dabbagh et al⁹ similarly described the management of CAD in a 64-year-old woman with dextrocardia using off-pump CABG. In that case, the primary operator stood at the left side of the patient, applying a RIMA graft to the LAD and saphenous vein grafts to the obtuse marginal and D1 arteries. Graft patency was confirmed at 2-year follow-up using computed tomographic angiography.

This report has discussed the management of a 56-yearold man with dextrocardia and morbid obesity who presented with severe diffuse, triple-vessel CAD with proximal LAD involvement. In addition to his unique anatomy, the patient was at high risk for both percutaneous and surgical revascularization because of several cardiac risk factors, including poorly managed type 2 diabetes, morbid obesity, and extensive smoking history. Given the patient's relatively young age, multivessel involvement, and unique anatomy, it was believed that his best chance for viable long-term revascularization would be to bypass his lesions using both arterial and venous grafts. As in previous reports of dextrocardia, the patient's LAD lesion was bypassed using RIMA instead of the left IMA (LIMA) as it would provide the most ideal conduit length and long-term patency. Because of the inversed orientation of the patient's heart, use of the LIMA would require excessive physical manipulation and stretching across the thorax, increasing the risk for damage during anastomosis as well as for premature graft failure.8 It was the surgeon's opinion that the length of the LIMA would not be enough to bypass the patient's LAD. Because of the patient's body habitus and excessive mediastinal and epicardial adipose tissue, a lateral pericardial window was created to bring the RIMA into the pericardium for anastomosis.

This report contributes to the literature describing the management of complex CAD in patients presenting with dextrocardia with situs inversus totalis. Satisfactory outcomes may still be achieved in patients with this anatomy and with several concurrent cardiovascular risk factors. As most heart team operators will have limited experience managing this unique anatomic variation, early multidisciplinary collaboration should be emphasized. Technical considerations, including careful conduit selection and sizing, operator positioning, and the optimization of pericardial windows for graft insertion, are also paramount. Finally, operators may consider use of the RIMA over the traditional LIMA for LAD le-

sions as well as open harvest of saphenous veins to increase venous graft quality for non-LAD lesions.

Article Information

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