

Mirror Mirror on the Wall- A Case of Dextrocardia with Situs Inversus

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ABSTRACT

Introduction: Dextrocardia with situs inversus is a rare congenital defect that causes reversal of the thoracic and abdominal organs and vessels. In regards to cardiac anatomy, the apex of the heart is right sided and the coronary ostia are reversed in patients with situs inversus and dextrocardia. Given the reversal of normal anatomic structures, successful coronary angiography requires several adjustments to engage the vessel and interpret the angiography.

Case report: This is a 67 year-old male with a history of dextrocardia with situs inversus and severe aortic stenosis who presented with dyspnea and orthopnea. Echocardiography confirmed critical aortic stenosis. He was referred for valve replacement and underwent coronary angiography. A non-standard counter-clockwise technique was required to engage right coronary artery and a double inversion technique was utilized to aid in interpretation of the angiogram.

Conclusion: Dextrocardia with situs inversus is a rare congenital defect and when not associated with Kartagener's carries a normal life expectancy. Therefore it is not unusual that patient's will need coronary angiography. Several technical modifications are needed for successful angiography.

Introduction

Dextrocardia with situs inversus is a rare congenital defect in which the apex of the heart is on the right side of the body. The incidence of dextrocardia with situs inversus is estimated to be around 1:8,000 to 1:10,000. The prevalence of coronary artery disease in patients with dextrocardia is similar to that in the general population. Coronary angiography in patients with dextrocardia presents several challenges including how to engage the vessels, as well as, angiographic interpretation. Here, we present a patient with dextrocardia and critical aortic stenosis that had coronary angiography performed using what has been described previously as the Double-Inversion technique. This technique was first described by Goel et al. and combines right to left reversal of the image during acquisition using the "horizontal sweep reserve" button on the machine plus a degree-to-degree reversal in the desired RAO/LAO angulations keeping the cranial/caudal tilts the same.

Case Report

The patient is a 67-year-old male with a history of dextrocardia situs inversus, severe aortic stenosis, and atrial fibrillation who

presented with dyspnea and orthopnea. Echocardiography confirmed a heavily calcified valve and severe aortic stenosis with a mean trans-valvular gradient of 52 mmHg, peak velocity of 4.5 m/s, dimensionless index of 0.19, and an aortic valve area of 0.68 cm². Given his symptomatic severe aortic stenosis, he was evaluated for valve replacement and underwent a preoperative coronary angiography. During angiography, his right coronary artery initially was difficult to engage with the standard clockwise rotation; however, it was successfully engaged with a counterclockwise rotation with a 6F JR4 catheter. The left main coronary artery, on the other hand, was easily engaged with a 6F JL4 catheter in a clockwise technique. Prone imaging format was used to record most images for easier interpretation. Coronary angiography demonstrated non-obstructive coronary artery disease, situs inversus, and right-sided aortic arch.

Discussion

Dextrocardia is a rare condition leading to the transposition of the heart in the right thoracic cavity with the apex pointing to the right side. When dextrocardia is not associated with Kartagener's syndrome, the condition carries a normal life expectancy. Therefore, it is not unusual for patients to live up to ages where senile aortic stenosis is encountered. The first cardiac catheterization in a patient with dextrocardia first occurred in 1973 by Hynes et al. . Since then, few have reported trans-femoral or trans-radial catheterization procedures.

Given the anatomic variance with reversed coronary ostia, several modifications are needed for successful angiography in patients with dextrocardia: proper catheter selection, catheter manipulation for successful cannulation, and mirror image angiographic angulation. Currently, there is no consensus on specific imaging catheters. Moreyra et al. reported that regular coronary catheters (Judkins) are difficult to engage the coronary ostia and recommend wider, nontraditional catheters. Others, specifically Kakouros et al. have reported that the coronary ostia can successfully be cannulated with Judkins catheters with opposite-direction catheter rotations (i.e. anticlockwise rotation in the ascending aorta for the right coronary artery). Mirror image angiographic angulation involves reversing the required right anterior oblique/left anterior oblique angles but keeping the cranial/caudal tilts the same. Other angiographers used a double-inversion technique, which normalizes all angiographic pictures to the standard conventional pictures as seen in a normally located heart using the "horizontal sweep reserve" button on the machine.

Keywords: Dextrocardia, Heart, situs inversus.