A Review of Situs Inversus and Dextrocardia

Ronnie O. Ortiz, MD, MSc; Murtuza J. Ali, MD; Fred Lopez, MD (Section Editor)

CASE REPORT

A 21-year-old woman at 25 weeks gestation presented to the emergency department with chief complaints of decreased appetite for one week, fever, runny and stuffy nose, and generalized muscle pains. She denied any previous medical or surgical history. Obstetrical history included spontaneous abortion at six weeks gestation without dilation and curettage. Her family history was negative for any medical problems. She denied ever using alcohol, tobacco or drugs. She led an active lifestyle and had no limitations on activity. She did not take any prescribed medications other than prenatal vitamins. Her initial vital signs at presentation revealed a blood pressure of 110/70 mmHg, heart rate 110-130 beats per minute, respirations 16-18 per minute, temperature 101.3 F, and oxygen saturation 99% on ambient room air. Her physical exam was reported as normal. She was given normal saline intravenously and oral acetaminophen with resolution of her fever and a subsequent decrease in heart rate to 100. Her hemoglobin was decreased at 9.9 mg/dL and the rest of her blood work and urine tests were normal. She was admitted to the obstetrical service for continued observation and fluid hydration.

On admission, an ECG was ordered due to her tachycardia (Figure 1). Due to ECG abnormalities, the cardiology department was consulted. A second ECG was done to ensure correct lead position and was unchanged, and was followed by a right sided ECG (Figure 2).

At time of the cardiology evaluation, her vital signs were normal. Her jugular venous pressure was normal. No carotid bruits were appreciated. Her heart sounds were distant and absent over the left chest but were normal in intensity on the right side of the chest. No murmurs, clicks or rubs were noted. No abdominal pain was appreciated with light palpation, but a limited abdominal exam was performed due to her gravid condition. Radial and dorsalis pedis pulses were intact and equal bilaterally. An echocardiogram confirmed suspicions for dextrocardia. Subsequently, she underwent further testing which confirmed situs inversus totalis.

DISCUSSION

The etymology of dextrocardia and DSIT are Latin, where dextro is derived from “dexter” meaning “right” and “cardia” meaning heart, when put together, “right-sided heart”. Situs in Latin is “position or site”, inversus totalis is “totally inversed”, hence totally inversed site, referring to the internal organs. Other variations include situs inversus incomplectus or situs inversus with levocardia where the heart remains on the left side.

Dextrocardia is defined as a congenital condition where the heart’s apex is oriented to the right side of the thorax, a mirror image of the heart, which is situated on the right side. In DSIT the major visceral organs and the heart are mirrored or reversed from their original position. These conditions are uncommon; dextrocardia occurs 1 in 12,000 births, DSIT occurs in approximately 1 in 10,000 births. Situs inversus incomplectus is the rarest form, occurring 1 in 2,000,000 live births.

DSIT is mostly associated with an autosomal recessive condition and less commonly X-linked. Associated congenital anomalies are seen in about 5% to 10% of cases, most commonly transposition of the great vessels; approximately 25% have primary ciliary dyskinesia. Conversely, 50% of those with primary ciliary dyskinesia have DSIT, a condition called Kartagener syndrome (triad of bronchiectasis, chronic sinusitis, and situs inversus). Cilia are the main determinants of organ orientation in vertebrate embryos and genetic mutations can alter and affect the cilia modifying their function leading or resulting in the respective position of the internal organs, right or left.

Patients with dextrocardia or DSIT without any other congenital anomaly or associated pathological condition live normal healthy lives. Randy Foye who plays for the Denver Nuggets in the National Basketball Association is an example of a patient with DSIT living a normal life. However, it is important for patients with this condition to relay this information to their physicians or health care providers as signs and symptoms of non-cardiac diseases
Figure 1: Abnormal findings include poor or no R wave progression across the precordium, abnormal right axis, and inverted P waves in lead I, aVL and upright aVR.

Figure 2: ECG with right-sided chest leads shows restoration of typical precordial R-wave progression. ECG= electrocardiogram, CT= computed tomography, MRI= magnetic resonance imaging, CXR= chest X-ray
such as appendicitis and cholecystitis may present with left sided pain, and cardiac chest pain may be right-sided in location. Also heart and especially liver transplantation surgeries may be challenging as placing a mirror image liver into an organ cavity is geometrical problematic and steps have to be taken to ensure proper anastomosis of the great vessels in the case of a heart transplant.

The diagnosis is relatively simple with an ECG or imaging study (CT, MRI, CXR, U/S, etc.). Commonly, dextrocardia and DSIT are diagnosed incidentally when people seek medical care for unrelated conditions, as occurred in this case. Typical ECG findings include precordial poor or no R wave progression, abnormal right axis with inverted P waves in lead I, aVL and upright aVR (Figure 1). These ECG findings may be confused with arm lead reversal. If there are no signs or symptoms and the heart is normal no tests or treatment are required. Signs and symptoms that are usually associated with congenital anomalies are more common in children and can be non-specific like shortness of breath, fatigue, acrocyanosis or cyanosis, frequent respiratory infections, failure to thrive, and jaundice.6

Dextrocardia and situs inversus are interesting from a diagnostic and embryological standpoint. These conditions, though uncommon, may be clinically important as they may hinder the diagnosis of certain diseases or acute conditions.

REFERENCES


Dr. Ortiz is a second-year fellow in the Department of Internal Medicine and Section of Cardiology at LSUHSC-New Orleans. Dr. Ali is an Associate Professor in the Department of Internal Medicine and Section of Cardiology at LSUHSC-New Orleans. Dr. Lopez is the Richard Vial Professor and Vice Chair in the Department of Medicine at LSUHSC-New Orleans.